Conclusions: The prevalence of the musculoskeletal manifestations is high (76.4%) in the hemodialysis population. Musculoskeletal disorders seem to cluster differently according to age and dialysis vintage.

Acknowledgements: Dr Ghada Abi Karam and Dr Fouad Fayad.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.4217

AB1337 CELIAC DISEASE AND RISK OF SARCOIDOSIS: A SYSTEMATIC REVIEW AND META-ANALYSIS

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Background: Several epidemiologic studies have demonstrated that patients with celiac disease may be at an increased risk of developing sarcoidosis. However, the data on this risk remain inconsistent.

Objectives: To compare the risk of developing sarcoidosis between patients with celiac disease and individuals without celiac disease.

Methods: A literature review was performed using MEDLINE and EMBASE database from inception to December 2017. Studies that compared the risk of sarcoidosis among patients with celiac disease versus those without celiac disease were included. The inclusion criteria were as follows: ¹ case-control, cross-sectional or cohort studies that investigated the risk of sarcoidosis among patients with celiac disease compared with individuals without celiac disease and ² odds ratios (OR), relative risks (RR), hazard ratios (HR) or standardised incidence ratios (SIR) with 95% confidence intervals (CI) or sufficient raw data to calculate these ratios were provided. Study eligibility was independently determined by 3 investigators. Differences in the determination of study eligibility were resolved by mutual consensus. Pooled odds ratio (OR) and 95% confidence interval (CI) were calculated using a random-effect, generic inverse variance method of DerSimonian and Laird.

Results: Of 375 retrieved studies, 4 studies (2 case-control studies and 2 cohort studies) with 6 93 639 participants met our eligibility criteria and were included in analysis. We found a higher risk of sarcoidosis among patients with celiac disease compared with individuals without celiac disease with the pooled OR of 7.16 (95% CI, 1.48–34.56). The statistical heterogeneity of this study was high ($I^2=95\%$). Funnel plot was relatively symmetric and did not suggest the presence of publication bias in favour of positive studies.

Abstract AB1337 – Figure 1. Forest plot of this meta-analysis

Conclusions: This systematic review and meta-analysis found a significantly higher risk of sarcoidosis among patients with celiac disease.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.4682

AB1338 COHORT OF PARAGUAYAN PATIENTS WITH EARLY ONSET ARTHRITIS

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Background: The PANLAR-EOA (early-onset-arthritis) project includes panamericanrheumatologists to determine regional characteristics of patients with early onset arthritis.

Objectives: To describe the cohort of Paraguayan patients included in PANLAR-EOA project.

Methods: Longitudinal, prospective, multicentric study. Patients were included according to the PANLAR-EOA project and registered in REPANARC (www.panlareoa.org) database. At baseline and annual visits, a large number of demographic, clinical and analytical variables were recorded. Quantitative variables were characterised by their means and standard deviations, while the qualitative

variables were characterised according to the percentage of patients. The comparison of epidemiological and clinical variables was performed using the chi-squared test and the Wilcoxon test respectively for qualitative and quantitative variables, respectively.

Results: 136 patients with early onset arthritis were included, out of which 88 completed the 12 months follow up and 58 the 24 months one. In these, 86% were female with a median age of 43.9±13.2 years. The most frequent race was mestiza in 80.1%. According to GRAFFAR index, middle class was the predominant social stratum (9.8±3.1). The average number of years of schooling was 12.8±3.8. Polyarticular onset was registered in 61% patients. During follow-up, 43.1% had positive rheumatoid factor and 56.5% positive anti-CCP. The diagnostic delay was 3.9±3.0 months. Initially, 63.2% (86/136) were diagnosed with rheumatoid arthritis (RA) and 36.8% (50/136) with undifferentiated arthritis (UA). The most frequent treatment was methotrexate (85.3%, 90.9%, and 89.3% at baseline, 1 and 2 years of follow-up respectively). During follow-up, a significant diagnostic change was observed in patients with UA ($p=0.004$, $OR=2.9$ [95%CI, 1.4–6.5]). The variables associated with RA diagnosis were presence of anti-CCP ($p=0.000$, $OR=15.8$ [95%CI, 5.4–51.1]), rheumatoid factor ($p=0.000$, $OR=9.2$ [95%CI, 3.4–27.0]), smoking ($p=0.032$, $OR=8.8$ [95%CI, 1.1–404.7]), high body mass index ($p=0.041$, $OR=1.94$ [95%CI, 0.2–4.1]) and high activity measured by the DAS28 index ($p=0.01$). After one year of follow-up there was a significant decrease in disease activity according to DAS 28 ($p=2.2e-09$ [95% CI, -1.5, -0.9]), SDAI ($p=1.2e-11$ [95% CI, 18.2, -11.2]) and HAQ ($p=7.2e-08$ [95% CI, -0.7, -0.4]). Similar results were found at the 2nd year of follow-up, DAS28 ($p=8.8e-06$ [95% CI, -1.6, -0.7]), SDAI ($p=2.1e-07$ [95% CI, -20.0, -10.3]) and HAQ ($p=3.4e-08$ [95% CI, -0.9, -0.5]).

Conclusions: In this cohort of early onset arthritis, diagnostic delay was lower than that observed in other series and the rate of change from diagnosis of UA to RA was statistically significant during the first year of follow-up. A good control of the inflammatory activity of the disease was observed, with a significant improvement of all the variables analysed during its evolution.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.6465

AB1339 ASSOCIATION OF ANTI-THYROID ANTIBODIES AND MUSCULOSKELETAL PAIN IN PATIENTS WITH AUTOIMMUNE THYROIDITIS

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Background: Autoimmune thyroiditis is closely associated with autoimmune diseases and can exhibit musculoskeletal pain as a clinical manifestation; the exact pathogenic mechanism of coexistence with other autoimmune disorders has not been clearly defined. There are very few studies that describe the association of anti-thyroid antibodies and musculoskeletal pain in patients with controlled autoimmune thyroiditis without rheumatologic disease

Objectives: To Determine the association of anti-thyroid antibodies and musculoskeletal pain in euthyroid patients with autoimmune thyroiditis who doesn't meet any criteria of a rheumatic disease

Methods: Fifty consecutive euthyroid (TSH test in a range of 0.34–5.6 μ U/mL and Ft4 0.54–1.64 ng/dL) patients with a diagnosis of autoimmune thyroiditis (positive anti peroxidase antibodies [TPOAb] ≥ 10 IU/mL and/or anti thyroglobulin antibodies [TGAb] ≥ 5 IU/mL), who attended the endocrinology clinic in an one-year period were included. At the moment of enrollment the patients didn't meet any classification criteria of a rheumatic disease. The presence of musculoskeletal pain was assessed using the survey "Program of modified community orientation for rheumatic diseases" (COPCORD) phase II, validated for the Mexican population, and used as a tool to determine the prevalence of rheumatic and musculoskeletal diseases [6]. COPCORD questionnaire consists of several sections such as number of affected joints, anatomic sites and pain severity measured by Visual Analogue Scale (VAS) with values ranged from 0 to 10. Descriptive statistical analyses were performed using the mean, range and standard deviation of variables. Correlations of statistical significance between groups were carried out using Spearman's rank correlation. P values < 0.05 were considered to be significant.

Results: 26 patients were TPOAb positive (830.76 IU/mL $\pm$ 1169.33 IU/mL); 41 patients were TGAb positive (257.126 IU/mL $\pm$ 629.344 IU/mL). There was a positive correlation between the presence of TPOAb with the number of affected joints (Spearman $r=0.9618$, $p=0.00001$) and with pain intensity (Spearman $r=0.9552$, $p=0.00001$). Similarly, there was a positive relationship between TgAb with the number of joints affected ($r=0.07800$, $p=0.00001$) and with pain intensity ($r=0.7268$, $p=0.00001$).

Correlation, (n)	Spearman <i>r</i>	<i>p</i> value
TPOAb versus intensity of pain, 26	0.9552	0.00001
TPOAb versus number of joints affected, 26	0.9618	0.00001
TPOAb versus age, 26	-0.3007	0.1328
TPOAb versus body mass index (BMI), 26	0.0024	0.9904
TgAb versus intensity of pain, 41	0.7268	0.00001
TgAb versus number of joints affected, 41	0.7800	0.00001
TgAb versus age, 41	-0.1615	0.6959
TgAb versus body mass index (BMI), 41	0.2172	0.1694

Abstract AB1339 – Figure 1

Conclusions: Our results suggests that in patients with controlled autoimmune thyroiditis, anti-thyroid antibody titers have a positive correlation with the presence of musculoskeletal pain and its severity; these findings deserve further study and research to establish the relationship of different autoantibodies present in autoimmune thyroid disease and musculoskeletal pain, as well as their role in rheumatic diseases, with a clinical importance not yet well established in this latter group.

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Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.6129

AB1340 DIFFERENTIAL CHARACTERISTICS IN A COHORT OF COLOMBIAN PATIENTS WITH THREE AUTOIMMUNE/AUTOINFLAMMATORY DISEASES: A NATIONAL REGISTRY UNDER A RISK MANAGEMENT MODEL

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Background: Latin American (LA) population with autoimmune/autoinflammatory (AID/AI) diseases is considered as a minority, with special characteristics of ancestry, socioeconomic status (SES) and cultural among others, with particular manifestations and prognosis outcomes

Objectives: To describe the largest cohort of patients with rheumatoid arthritis (RA), spondyloarthropathies (SpA) and systemic lupus erythematosus (SLE), in Colombia, evaluated through the social health care security system (SHCSS) and under a risk management model highlighting shared characteristics, as well as differences

Methods: A national register-based retrospective cohort study of the SHCSS since 2015 assessing three AID/AI diseases (RA, SLE, SpA) treated in a centre of excellence in rheumatology, under a risk management program in six cities of Colombia. Data about covered population was obtained from the integral information system of SHCSS (SISPRO). Clinical records of patients with AID/AI ICD-10 codes were selected. Adult patients fulfilling international validated diagnoses criteria for the three AID/AI were included. A survey data collection instrument was implemented. Analyses (SAS software V9.4) with standard statistical methods (Polytomous logistic regression, Chi-squared test Fisher's exact test, and Kruskal-Wallis test) were done. Odds ratio (OR) and 95% percent CIs were computed ($p < 0.05$)

Results: Of 1 62 487 population covered from national security system (SISPRO data) having AID/AI diseases, 71 515 were evaluated in the 6 cities and 12 323 records were selected of patients by population census attended in the institution. A total of 5372 records meet international validated criteria and were included (RA=3576, SLE=1175, SpA=621). RA patients were older, had lower educational level and SES, longer duration of the disease and late age at onset. Cardiovascular compromise was high in RA and SLE. Polyautoimmunity was predominant in SLE. A total of 89% of patients were under steroids treatment (SLE and RA) and 35% in SpA. Biologics were indicated in 59% (SpA), 23% (RA) and 7,1% (SLE). EuroQol five dimensions scores were significantly better in the SLE group (time of admission). Additional clinical characteristics are described in Table 1

Abstract AB1340 – Table 1. Main Clinical characteristics in the three groups

Clinical compromise	RA n=3576		SLE n=1175		SpA n=621	
	N	%	N	%	N	%
Cardiovascular*	1397	39,1	371	31,6	13	2,1
Pulmonary *	112	3,1	65	5,5	6	0,96
Central and peripheral nervous system*	67	1,8	63	5,4	5	0,8
Cutaneous*	170	4,7	881	75	298	47,9
Ocular*	415	11,6	130	11,1	112	18
Haematological*	90	2,5	970	82,5	10	1,6
Hepatic*	141	3,9	54	4,6	51	8,2
Renal*	11	0,3	473	40,2	4	0,64
Polyautoimmunity**	137	3,8	248	21,1	15	2,4

*p-value<0.0001 ** p-value<0,0001

Conclusions: This is the largest cohort evaluated through the SHCSS and under a risk management model in Colombia (LA). This population share special characteristics, highlighting differences between these three AID/AI diseases that are frequently evaluated in the same scenario in real life conditions by the rheumatologist. It is worth noting the specific poor prognosis factors of RA and the high percentage of patients under biological treatment in SpA group, which implies a high cost for the health system

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Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.5764

AB1341 PROFILE AND IMPORTANT CHARACTERISTICS OF RHEUMATOLOGICAL DISEASES MANAGED OVER ONE YEAR IN A URBAN UNIVERSITY BASED RHEUMATOLOGY CLINIC IN KUALA LUMPUR, MALAYSIA

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Background: The Rheumatology Unit of University Malaya in Kuala Lumpur is located in the University Malaya Medical Centre which is a strategically placed 1008 bed, tertiary healthcare institution. An audit of the rheumatology patients was carried out in 2017.

Objectives: The main objective is to create a database and registry of all the cases that are managed as outpatient over a period of 1 year. The data is analysed to provide the required information Details regarding therapy with biologics and small molecules are collected and analysed.

Methods: A case report form was designed for each patient, which include fields capturing first time visit and subsequent visits. Information captured were demographic data, disease diagnosis and duration, clinical features, laboratory values, co-morbidity and current medications. Longitudinal data was captured. All information were entered into RedCap software with an institutional license. SPSS statistical analysis software was used.

Results: Total number of patients in 2017 was 2329. The total number of patient visits was 8255. There was preponderance of females (1,856, 79.7%) compared to males (473, 20.3%). For ethnicity, Chinese patients constitute the most (1,041, 44.5%), followed by Malay (697, 29.8%), Indian (565, 24.2%) and others ethnicities (34, 1.5%). In this population, 3 most common rheumatological diseases seen are Rheumatoid arthritis (760, 32.6%), osteoarthritis (610, 26.2%) and Systemic Lupus Erythematosus (514, 22.1%). Following in descending order are gout (158, 6.8%), soft tissue rheumatism (153, 6.6%), psoriatic arthritis (118, 5.1%), Sjogren syndrome (102, 4.4%), scleroderma (58, 2.5%), mixed connective tissue diseases (49, 2.1%), vasculitis (44, 1.9%), ankylosing spondylitis (36, 1.5%), overlap syndrome (29, 1.2%), Adult onset Still's disease (26, 1.1%), dermatomyositis/polymyositis (25, 1.1%), and fibromyalgia (20, 0.9%). Most recent management activity after clinical assessment, majority of patients (1391, 60.8%) were noted to have no change in management (same therapy continued, but not in remission), followed by those in remission (544, 23.6%). 215 (9.3%) patients require escalation therapy. 13 patients (0.6%) required hospitalisation. 112 (4.9%) had de-escalation therapy. There were 13 deaths (0.6%). Most patients on biologics or small molecules are diagnosed with either Rheumatoid arthritis, SLE, Ankylosing Spondylitis and Psoriatic Arthritis. 44.7% of the patients on biologics have moderate disease activity while 33% of them have mild disease activity. 13.8% of them had achieved remission and 8.5% of them continued to have severe disease activity. The overall assessment of disease activity noted that 57 (2.5%) had severe

disease activity, 550 (23.9%) had moderate disease activity, 1139 (49.5%) considered as mild disease activity and 557 (24.2%) were considered to be in remission.

Conclusions: This study has created a database and registry from which important data has been derived. Majority of the patients are being managed sub-optimally, most likely due to limited access to advanced therapies in prominent rheumatological diseases that are intrinsically active and respond modestly to conventional DMARDs and therapy. Further analysis is ongoing

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.6326

AB1342 RHEUMATIC DISEASES PREVALENCE AND QUALITY OF LIFE IN SARAGURO INDIGENOUS POPULATIONS OF ECUADOR: A CROSS-SECTIONAL COMMUNITY-BASED STUDY

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Background: Rheumatic diseases are more prevalent and aggressive in indigenous population groups, in which providing medical attention poses a challenge for the rheumatologist.

Objectives: To estimate the prevalence of musculoskeletal (MSK) disorders and rheumatic diseases in the Saraguro indigenous people and their impact on the quality of life.

Methods: Cross-sectional analytical study carried out in the community of Saraguro using the COPCORD methodology. Mixed and randomised sampling techniques were employed. The following validated questionnaires were administered: 1. Screening for musculoskeletal (MSK) disorders and rheumatic diseases. 2. A sociodemographic questionnaire. 3. A functional capacity questionnaire (HAQ-DI) and an instrument to measure workload and repetitive movements. 4. Quality of life (EQ-5D3L). Cases with MSK disorders were reviewed by rheumatologists within the community.

Results: A total of 2687 individuals over 18 years of age participated, with an average age of 44 (SD 19.9) years; 1690 (62.9%) were women, 872 (32.4%) were Kwichua speakers; 2108 (78.4%) were employed, of these 32.5% were farm workers. MSK pain was reported in 1244 (46.3%); pain was severe in 448 (36%); 868 (69.7%) used some medical treatment and 1013 (81.4%) used traditional medicine. The most prevalent self-reported comorbidities were anxiety (55.5%) and depression (46.8%). Rheumatic diseases were diagnosed in 861/1244 (69.2%), with the following most prevalent conditions: Low back pain 9.3%, hand osteoarthritis 7.2%, knee osteoarthritis 6.5%, RRPS (rheumatic regional pain syndrome) 5.8%, fibromyalgia 1.8%, rheumatoid arthritis (RA) 1.3%. Disability (HAQ >0.8) was observed in 356 (28.6%), whereas loading and pushing objects heavier than 20 kg and shaking hands was significantly associated with MSK pain. The regression models showed a significant association with a lower quality of life in those with lower education levels (OR=0.89; 95% CI 0.88 to 0.91, p<0.001), physically demanding jobs (OR=0.54; 95% CI 0.42 to 0.69, p<0.001), cooking with firewood (OR=1.55; 95% CI 1.05 to 2.30, p=0.02), high blood pressure (OR=2.31; 95% CI 1.73 to 3.10, p<0.001), and rheumatic diseases, especially RA (OR=5.52; 95% CI 2.48 to 12.27, p<0.001) and hands OA (OR=2.05; 95% CI 1.44 to 2.91, p<0.001). One finding is that having RA was associated with cooking with firewood, and that smoking was almost nonexistent in this population. This suggests that wood smoke pollutants play a similar role to that described with tobacco smoke.

Conclusions: A high prevalence of MSK disorders, rheumatic diseases and RA was found. The prevalence of rheumatic diseases was associated with a lower education level, cooking with firewood, and physically demanding jobs. The greatest impact on the quality of life in all dimensions was on the individuals with RA and hand OA.

Acknowledgements: Funding: Research Dept. Universidad de Cuenca, Ecuador

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.4857

AB1343 PREVALENCE OF AUTOIMMUNE DISEASES IN CATALONIA: A POPULATION BASED STUDY USING A PUBLIC BIG DATA ANALYTICS (PADRIS)

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Objectives: To analyse the prevalence of autoimmune diseases (ADs) in Catalonia by using a public big data program (Public Data Analysis for Health Research and Innovation Program, PADRIS)

Methods: We used the health insurance database of the Catalan National Health Insurance (CNHI) which includes all catalan population registered as insured population in 2016. The sample included 7,483,761 inhabitants. ADs were identified according to the corresponding International Classification of Diseases, Ninth Revision, Clinical Modification (ICD-9-CM) codes. A total of 33 autoimmune diseases were analysed classified in 4 main categories: rheumatic, systemic, organ-specific and immunodeficiency/autoinflammatory. The prevalence of ADs was calculated as the number of ADs patients divided by the total CNHI beneficiaries in the same year (rate per 1 00 000 persons, 95% confidence intervals -CI-)

Results: The overall prevalence of ADs was 1202 per 1 00 000 persons (95% CI 1,194–1,209); the prevalence was 1455 (95%CI 1,443–1,467) in women and 939 (95%CI 929–949) in men. ADs were classified as organ-specific (43%), systemic (33%), rheumatic (23%) and immunodeficiency/autoinflammatory (1%) autoimmune diseases. The ADs with the highest prevalence rates included psoriasis (282 cases per 100,000, 95% CI 278–286), rheumatoid arthritis (RA) (178 cases per 100,000, 95% CI 175–181), polymyalgia rheumatic (98 cases per 100,000, 95% CI 96–101), spondyloarthropathies (92 cases per 100,000, 95% CI 90–94), systemic lupus erythematosus (SLE) (68 cases per 100,000, 95% CI 66–70) and Sjögren's syndrome (59 cases per 100,000, 95% CI 57–61). In 26 (79%) of the 33 ADs, the female:male ratio was higher than 1; the highest rates were reported for Sjögren's syndrome (10.5:1), primary biliary cholangitis (5.8:1), SLE (5.4:1), systemic sclerosis (3.4:1) and rheumatoid arthritis (2.6:1). An enhanced prevalence of ADs was reported in Southern regions (1225 cases per 1 00 000 persons in Barcelona/Tarragona regions -CI95% 1,216–1,233- vs 1075 cases per 1 00 000 persons in Girona/Lleida regions -CI95% 1,056–1,093-, p<0.001).

Conclusions: Nearly 90 000 catalans are classified as having an autoimmune disease, representing a prevalence of 1.2% of the total catalan population, a rate which reaches 1.5% in women. The highest frequencies are reported for psoriasis (0.28%), rheumatoid arthritis (0.18%), polymyalgia rheumatica (0.10%), spondyloarthropathies (0.09%), lupus (0.07%) and Sjögren syndrome (0.06%).

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.5974

AB1344 PRESENCE OF ECHOCARDIOGRAPHIC CRITERIA FOR HFPEF MULTIPLIES THE RISK FOR DEATH AND CARDIOVASCULAR EVENTS IN PATIENTS WITH RHEUMATIC DISEASES

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Background: Patients with rheumatic diseases (RD) have an increased risk for cardiovascular (CV) disease and heart failure (HF). Clinical assessment of HF signs and symptoms in RD is often limited by functional impairment.

Objectives: We investigated the prognostic value of echocardiographic and neurohormonal criteria for HF with preserved ejection fraction (HFpEF) in patients with RD.

Methods: This prospective, single-centre study included consecutive RD outpatients considered at increased risk for CV events according to ESC score (>3%), pathological ECG, or elevated NTproBNP (>200 pg/mL) as published by this group*. Clinical assessment and transthoracic echocardiography according to

ESC criteria was performed. Presence of HFpEF was assumed if patients had NTproBNP >125 pg/mL, and either left ventricular hypertrophy (LVH; averaged septal-posterior wall thickness >11 mm) or left atrial dilation (LAD; LA Diameter >43 mm), regardless of presence of dyspnea. Kaplan-Meier plots were generated, and hazard ratios (HR) with 95% confidence intervals were computed using Cox regression with adjustment for age.

Results: Out of 764 patients (mean age 51 years, 70% female) 46% had rheumatoid arthritis (RA), 34% systemic autoimmune diseases (SAI; connective tissue disease or vasculitis), and 20% spondylo-arthritis (SpA); 248 of these patients (mean age 61±13 years, 62% female, RA 46%; SAI 32%; SpA 22%) had valid echocardiographic data (4 patients with LV ejection fraction <50% were excluded; follow-up data was missing in 3 cases). After a median follow-up time of 5.4 years, 20.6% of patients (group 1–4: n=19/5/8/1 respectively) had died or suffered a CV event (myocardial infarction 4.1%; stroke 1.8%; decompensated HF 1.8%; resuscitation 0.9%).

In univariable analysis NTproBNP >125 pg/ml (HR 3.6; 1.9–6.8, $p=0.0001$), LAD or LVH (HR 2.3; 1.1–4.5, $p=0.02$), and age per 5 years (HR 1.4, 1.2–1.6, $p<0.0001$) were significant predictors for an increased risk for death or CV event. Compared to the referent group consisting of patients with no signs of LVH or LAD in the presence of normal NTproBNP (group 4; n=59, 23.8%), patients with echocardiographic criteria for HFpEF (group 1; n=84, 33.9%) had a 7-fold increased risk for death or CV event: HR 7.2 (1.6–31.9; figure). The event risk for both patients with elevated NTproBNP but absent LVH or LAD (group 2; n=49, 19.8%) as patients with normal NTproBNP but presence of LVH or LAD (group 3; n=56, 22.6%) was also 5- to 6-fold increased: HR 5.5 (1.2–25.8) and 5.5 (0.9–18.2), respectively.

Conclusions: In patients with RD with an increased baseline CV risk, echocardiographic criteria suggestive of HFpEF are highly relevant indicators of worse outcome (7-fold increased risk for death or CVE), in particular in conjunction with an elevated NTproBNP value, irrespective of clinical presentation.

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Acknowledgements: This study was supported by the Competence Network Heart Failure Germany (BMBF grant 01 GI0205/01GI1202A) and the Comprehensive Heart Failure Centre Würzburg (BMBF grant 01EO1004).

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.4875

AB1345

EVALUATION OF MACROPHAGE ACTIVATION SYNDROME IN HOSPITALISED FEBRILE PATIENTS WITH KIKUCHI-FUJIMOTO DISEASE BASED ON THE 2016 EULAR/ACR/PRINTO CLASSIFICATION CRITERIA

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Background: The incidence and the risk of MAS in Kikuchi-Fujimoto disease (KFD) have been poorly understood, probably because of the rarity of the disease.

Objectives: To evaluate the 2016 classification criteria for MAS in hospitalised febrile patients with KFD and compare the clinical features of MAS in KFD with those of systemic lupus erythematosus (SLE) and adult-onset Still's disease (AOSD).

Methods: The records of febrile patients with KFD, hospitalised between November 2005 and April 2017, were reviewed. Patients were evaluated for MAS using the 2016 classification criteria. Clinical and laboratory features between patients with KFD with and without MAS, as well as those of patients with MAS in KFD, AOSD, and SLE were compared.

Results: Among 78 patients hospitalised with KFD, 24 had MAS during admission. Patients with MAS had longer hospital stays than patients without MAS and only patients with MAS required intensive unit care and experienced in-hospital mortality. Patients with MAS had higher C-reactive protein, aspartate aminotransferase, alanine aminotransferase, lactate dehydrogenase, and ferritin levels and lower lymphocyte count, platelet count, and total protein and albumin levels. MAS patients with KFD, AOSD, and SLE exhibited different clinical characteristics

according to the underlying disease; however, MAS patients with KFD required less intensive immunosuppressive treatment than those with AOSD and SLE.

Abstract AB1345 – Table 2. Comparison of baseline laboratory findings of patients with MAS and without MAS

Variable	Without MAS (n=54)	With MAS (n=24)	p-value
General laboratory variables			
WBC count (/μL)	3475.0 (2350.0–5000.0)	3285.0 (2335.0–5905.0)	0.996
Haemoglobin (g/dL)	12.5 (11.7–13.4)	11.7 (10.3–13.2)	0.102
Neutrophil count (/μL)	1925.0 (1310.0–3050.0)	2206.5 (1375.0–4833.5)	0.378
Lymphocyte count (/μL)	1045.0 (860.0–1370.0)	600.0 (490.0–940.0)	<0.001
ESR (mm/h)	48.5 (27.0–64.0)	52.5 (39.5–72.5)	0.242
CRP (mg/L)	15.0 (7.5–37.1)	52.6 (27.7–97.1)	<0.001
Creatinine (mg/dL)	0.7 (0.6–0.8)	0.7 (0.5–1.1)	0.349
ALT (IU/L)	21.5 (14.0–38.0)	44.5 (23.0–153.0)	0.003
Albumin (mg/dL)	4.0 (3.7–4.3)	3.4 (3.1–3.7)	<0.001
LDH (IU/L)	439.5 (291.0–611.0)	771.5 (460.0–1213.5)	0.003
Laboratory variables selected for MAS classification			
Platelet count (× 1000/μL)	195.5 (155.0–260.0)	139.0 (94.5–184.0)	<0.001
AST (IU/L)	29.0 (22.0–39.0)	93.5 (62.5–191.0)	<0.001
Ferritin (ng/mL)	242.8 (112.0–405.2)	1449.7 (812.9–4463.1)	<0.001
Triglyceride (mg/dL) [†]	89.0 (60.0–106.5)	133.0 (90.3–161.3)	0.076
Fibrinogen (mg/dL) [§]	342.0 (338.0–346.0)	317.5 (227.5–401.0)	0.695
Bone marrow biopsy findings*			
Presence of hemophagocytosis	0/7 (0.0)	2/12 (16.7)	0.509

Data are expressed as median (interquartile range) or n (%).

ALT, alanine aminotransferase; AST, aspartate aminotransferase; CRP, C-reactive protein; ESR, erythrocyte sedimentation rate; LDH, lactate dehydrogenase; MAS, macrophage activation syndrome; WBC, white blood cell

Abstract AB1345 – Figure 1. Comparison of HScore and the probability of having hemophagocytic syndrome in patients with KFD with and without MAS

Comparison of HScore (A) and the probability of having hemophagocytic syndrome (B) in KFD patients with MAS and without MAS

HScore, hemophagocytic syndrome score; KFD, Kikuchi-Fujimoto disease; MAS, macrophage activation syndrome.

Conclusions: The 2016 classification criteria were useful in identifying KFD patients with poor clinical outcome. Although treatment for patients with MAS in KFD, AOSD, and SLE differed according to the underlying disease, the 2016 classification criteria for MAS might aid in stratifying high-risk patients and provide guidance for treatment.

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Acknowledgements: None.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.5007