Methods: Four regional hospitals participated with study period from 1/1/2006 to 30/6/2016. IgG4RD patients were classified into definite, probable and possible IgG4RD according to Japanese Comprehensive Diagnostic Criteria for IgG4RD (3).

Statistical analysis: Association between the individual categorical covariates and the different organ involvement were analyzed by Fisher's exact test.

Results: 108 patients were included. There were 81 male patients and 27 female patients and the male to female ratio is 3:1. The mean age of diagnosis was 62.8 year old. 57 patients were diagnosed as definite IgG4RD (53%), 14 patients as probable IgG4RD (13%) and 37 patients as possible IgG4RD (34%). Salivary glands involvement was the commonest, (M=45.7% vs F=33.3%), followed by pancreas (M=28.4% vs F=18.5%) and biliary system in male (M=24.7% vs F=11.1%) and orbital in female (M=23.5% vs F=18.5%). 53% patients also had multi organ involvement.

104 patients had serum IgG4 level checked and it was raised in 94 patients (90.4%). 77 patients (74.0%) had IgG4 level above twice upper limit of normal. IgG4 level was normal in 10 patients. 98% and 86.3% patients with multi organ involvement had IgG4 level >135mg/dL (p=0.016) and >270mg/dL (p=0.007) respectively. In the hepatobiliary group (n=36), 30 patients had blood checked for Ca19.9 level and it was raised in 17 patients (56.7%), with the highest level 14127 U/ml. 71 histological reports were available. Lymphoplasmacytic infiltration was a consistent features in all specimens (100%). Eosinophilic infiltration was seen in 19 specimen (27%). Fibrosis was seen in 44 specimens (63%). Sialadenitis and dacryoadenitis were associated with dense fibrosis. Obliterative phlebitis was seen in 7 specimens (10%). IgG4/IgG ratio was reported in 61 specimens. 53 specimens had IgG4/IgG ratio greater than 40% per high power field which accounted for 86.9% of the specimens.

Conclusions: Most patients were male with the peak diagnosis at age 50 to 70. Salivary gland and pancreas were the most common organ involved. Most patients had a raised IgG4 level and most histological specimens showed a raised IgG4/IgG ratio on immunostaining. IgG4 level twice the upper limit of normal was quite specific for the disease and was associated with multi organ involvement.

References:

- [1] Pieringer H, et al. IgG4- related disease: an orphan disease with many faces. *Orphanet journal of rare diseases*. 2014;9:110.
- [2] Haitao Du YW et al. IgG4-related disease and the currency status of diagnostic approaches. *EXCLI*. 2012;11:651–8.
- [3] Umehara H, et al. Comprehensive diagnostic criteria for IgG4-related disease (IgG4-RD). 2011. *Modern Rheumatology*. 2012;22(1):21–30.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2017-eular.2145

FRI0600 THE EFFICACY OF DIFFERENT COLCHICINE DOSES IN TREATMENT OF FAMILIAL MEDITERRANEAN FEVER PATIENTS

E. Uğuztemur, S. Çelik, S. Sadri, E. Aşçı, G. Ergun, Ö. Velipaşalar, S. Yılmaz-Öner, C. Bes, N. Alpay-Kantantz. *Bakırköy Dr.Sadi Konuk Research and Training Hospital, Istanbul, Turkey*

Background: Recommended colchicine dose in familial mediterranean fever (FMF) is 1–1.5 mg/day in adults. If necessary it can be increased up to 3 mg/day by clinical monitoring. In the final FMF recommendation report of the EULAR, it was reported that after 5 years of stable disease, in appropriate patients, the dose of colchicine may be reduced by close follow-up. However, the number of clinical trials supporting this recommendation is not enough.

Objectives: In this study, demographic and clinical characteristics of FMF patients whose disease under control and treated with different doses of colchicine were compared.

Methods: Among the all FMF patients attending to the rheumatology outpatient clinic between April and September 2016, only the patients in non-attacks period at least for the last 1 month were included to the study. Clinical and laboratory findings, MEFV gene mutations, drug compliance of the patients and duration and doses of the colchicine treatment were recorded. The 'international severity scoring system for familial mediterranean fever' (ISSF) score was calculated to determine disease severity. The patients who were under the control (less than 4 attacks per year and ISSF score ≤2) divided to three groups. These groups were the patients taking low dose (average daily dose of colchicine <1 mg), median dose >1 mg/day and <1.5 mg/ day and standard dose ≥1.5 mg/colchicine treatment at least for 3 years. Patients treated with DMARDs or biological therapy were excluded from the study.

Results: Of the 162 FMF patients enrolled into the ongoing cohort, 86 patients were excluded from the study due to 57 patients had active disease, 19 had not yet completed 3 years of colchicine treatment and 10 patients received biological or DMARD treatment. The remaining 76 patients (25 male, 51 female) met the study criteria. The mean age was 32±11.5 years and the disease duration was 18.2±10.9 years. Amyloidosis or organ failure was not detected in any of these patients. Because of various reasons (drug side effect, treatment incompatibility, physician advice), 16 patients (21%) were treated with low dose colchicine (group A), 30 patients (39%) with moderate dose of colchicine (group B) and the remaining 30 patients (39%) with standard dose colchicine (group C). There were no statistically significant differences in demographic, clinical or laboratory data among these 3 subgroups.

Conclusions: Disease control in FMF is important to prevent amyloidosis and improve the quality of life. Recent studies contribute to the determination of treatment goals. Reducing the colchicine dose may increase drug compliance in this lifelong treatment. In our study, there was no difference between the treatment groups in amyloidosis, MEFV gene mutation or subclinical inflammation. These findings suggest that the dose of colchicine maybe reduced in inactive patients who determined by the number of annual episodes and the ISSF score.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2017-eular.6885

FRI0601 HYPERFERRITINEMIC SYNDROME IN A GENERAL UNIVERSITY HOSPITAL

F. Pierini, I.J. Gandino, J.M. Martinez Perez, S. Ruta, M. Scolnik, E.R. Soriano. *Rheumatology Section, Medical Services, Hospital Italiano de Buenos Aires, Buenos Aires, Argentina*

Background: Hyperferritinemia is associated with severe inflammatory conditions, such as rheumatic diseases with systemic inflammatory responses, and multiorgan dysfunction syndromes.

Objectives: To determine which diseases are associated with hyperferritinemia in a tertiary hospital; to compare ferritin levels between these different entities and to evaluate relationship between levels of ferritin and mortality in these patients.

Methods: A retrospective study was carried out in which all patients over 18 years with at least one determination of serum ferritin equal to or greater than 1000 ng/ml were identified in the laboratory database of our hospital between 1/1/2006 and 6/30/2016. Corresponding electronic medical records were reviewed and demographic data and clinical data were collected. Mortality was assessed at the end of follow-up. Descriptive statistical analysis and logistic regression analysis were performed in order to identify variables associated with mortality.

Results: A total of 1979 patients were included, 1235 men (62.4%) with a mean age of 63.2 years (SD 17.2). Only 36 patients (1.8%) presented a rheumatologic diagnosis as the only cause of high levels of ferritin, with Still's disease (n=8) and systemic Vasculitis (n=9) being the main diagnoses. Table 1 shows patients' characteristics grouped according to whether the elevation of ferritin was associated with a rheumatic disease or not. Median serum ferritin and transferrin saturation in both groups were similar (see Table 1). Mortality was lower for rheumatologic causes (5.9% vs 37.2%, p<0.001). Variables that were associated with mortality in multivariable logistic regression analysis were: maximum ferritin value (OR 1.0004, 95% CI 1.0003–1.0004, p<0.001) and age (OR 1.03, 95% CI 1.02–1.04, p<0.001), whereas the diagnosis of a rheumatic disease was a protective factor (OR 0.11, 95% CI 0.03–0.47, p=0.003). ROC curve for ferritin and mortality showed an area under the curve of 0.59 (95% CI 0.58–0.62). Ferritin levels greater than 3000 ng/ml showed a specificity of 89.2% and a sensitivity of 19.7% for mortality, regardless of cause of ferritin elevation.

Table 1. Patients' characteristics grouped by cause of Hyperferritinemia (Rheumatologic or not)

	Rheumatic disease (n=35)	Other causes of ferritin elevation (n=1944)	p
Female, n (%)	13 (37.1)	731 (37.6)	0.96
Mean age, years (DS)	62.5 (17.1)	52.4 (21.1)	<0.001
Diagnostic			
Vasculitis	9	Solid cancer	497
Still's disease	8	Infections	336
Rheumatoid arthritis	4	Onco-hematologic disease	302
SLE	3	Chronic renal insufficiency	236
Seronegative arthritis	3	Hepatic disease	188
and psoriatic arthritis	3	Others	152
Others	3	Hematologic disease	128
Gout	2	Cardiovascular	57
Juvenile idiopathic arthritis	1		
Myositis	1	Sepsis	46
IgG 4	1	Iron overload	2
Serum ferritin, median (IQR)	1622 (1264- 3639)	1460 (1200–2140)	0.07
Transferrin saturation, %, median (IQR)	38 (19–50)	33 (18–55)	0.83
Follow-up time, years, median (IQR)	5.2 (1.2–10.2)	5.7 (1.2–8.9)	0.97
Mortality, n (%)	2 (5.9)	698 (37.2)	<0.001

Conclusions: Rheumatic diseases, represents a very small percentage of the causes of elevation of ferritin above 1000 ng/ml, and were associated with lower mortality than the non-rheumatic causes. Serum ferritin levels were significantly associated with increased mortality regardless of the underlying cause.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2017-eular.2460

FRI0602 NATIONAL RECOMMENDATIONS ON THE USE OF IMMUNOMODULATORY DRUGS IN PATIENTS WITH NON-INFECTIOUS NON-MALIGNANT ANTERIOR UVEITIS

G. Espinosa¹, S. Muñoz-Fernandez², J.M. García Ruiz de Morales³,

J.M. Herreras⁴, M. Cordero-Coma⁵. ¹Department of Autoimmune Diseases, Institut Clinic de Medicina i Dermatologia, Hospital Clinic, Barcelona; ²Rheumatology, Hospital Universitario Infanta Sofia, Madrid; ³Immunology, Complejo Asistencial Universitario de León, León; ⁴Ophthalmology, Hospital Clínico Universitario de Valladolid, Valladolid; ⁵Ophthalmology, Complejo Asistencial Universitario de León, León, Spain

Background: Anterior uveitis (AU) is the most common pattern of uveitis, that might lead to important ocular complications including blindness. Immunomodulatory drugs have been used in order to prevent recurrences of uveitis. Nevertheless, it is not clear which drug could be preferred in each patient.

Objectives: To generate recommendations on the use of immunomodulators in adult patients with non-infectious, non-neoplastic anterior uveitis (AU) based on best evidence and experience.

Methods: Delphi methodology was followed. A multidisciplinary panel of 5 experts (2 ophthalmologists, one immunologist, one rheumatologist, one internist) was established, who, in the first nominal group meeting, defined the scope, users, and chapters of the document. A systematic literature review was performed to assess the efficacy and safety of immunomodulators in patients with non-infectious, non-neoplastic AU. All of the exposed above was discussed in a second nominal group meeting and 33 recommendations were generated. Recommendations agreement grade was tested also in 25 additional experts. Recommendations were voted from 1 (total disagreement) to 10 (total agreement). We defined agreement if at least 70% voted ≥ 7 . The level of evidence and grade or recommendation was assessed using the Oxford Centre for Evidence-based Medicine Levels of Evidence.

Results: The 33 recommendations were accepted. They include specific recommendations on patients with non-infectious, non-neoplastic UA, as well as different treatment lines. Methotrexate (MTX) or Sulfasalazine were recommended as a first line drugs in refractory cases to topic treatments in patients with AU and spondyloarthritis, inflammatory bowel disease, psoriasis, idiopathic HLA-B27 positive or negative AU. Etanercept was recommended for patients with TRAPS syndrome, and for those with other autoinflammatory syndromes canakinumab or anakinra. In case of bilateral sarcoidosis, relapsing polychondritis or TINU syndrome, MTX was recommended along with systemic steroids. For patients with a flare of AU and Behçet disease, systemic steroids along with azathioprine or a calcineurin inhibitor were recommended. The indication of an immunomodulatory drug in patines with multiple sclerosis was considered to be decided with a neurologist. For patients refractory to all exposed above and/or intolerant, depending on AU type, a change to another classical immunomodulatory drug or to an anti-TNF was recommended (adalimumab, infliximab, certolizumab or). Except for patients with TINU, etanercept was not recommended because current evidence does not support the use of it to prevent AU flares.

Conclusions: In patients with non-infectious, non-neoplastic AU, these recommendations on the use of immunomodulators might be a guide in order to help in the treatment decision making, due to the lack of robust evidence or other globally accepted algorithms.

Disclosure of Interest: G. Espinosa Grant/research support from: GSK, Actelion, S. Muñoz-Fernandez: None declared, J. García Ruiz de Morales: None declared, J. Herreras Grant/research support from: Allergan and Abbvie, M. Cordero-Coma Grant/research support from: Abbvie, Merck Sharp & Dohme, and Allergan
DOI: 10.1136/annrhumdis-2017-eular.4810

FRI0603 CLINICAL IMPLICATIONS OF ULTRASONOGRAPHY IN MONITORING DISEASE ACTIVITY OF RELAPSING POLYCHONDRIITIS: THE ASSESSMENT OF AURICULAR CHONDRIITIS IN 3 CASES

H. Nishikawa, Y. Taniguchi, Y. Terada. Kochi Medical School, Nankoku, Japan

Background: Relapsing polychondritis (RP) is a rare systemic inflammatory disorder and might often be refractory. Therefore, the discover of imaging modality would be required on diagnosis and treatment of RP.

Objectives: To assess the clinical implications of ultrasonography (US) in monitoring disease activity of relapsing polychondritis (RP).

Abstract FRI0604 – Table 1. Patient demographic and clinical characteristics

Patient	1	2	3
Age, years	71	65	46
Sex	Male	Male	Female
Race	Caucasian	Caucasian	Caucasian
Tumor	Metastatic melanoma	Metastatic melanoma	Metastatic melanoma
CPI	Anti-CTLA4 antibody	Anti-PD-1 antibody	Anti-PD-1 antibody
Onset of arthritis-irAEs (days after initiation of CPI)	100	19	49
Pattern of arthritis	Symmetric polyarthritis involving small joints	Symmetric polyarthritis involving small and large joints	Symmetric polyarthritis involving small joints
History of autoimmune disease prior to CPI treatment	No	No	No, but known positive ANA
Autoantibody results	ANA, CCP neg RF: 27 (UNL: 15.9)	ANA, CCP, RF neg	CCP, RF, Ro, dsDNA neg ANA 1:640 (homogenous) La pos
Discontinuation of CPI treatment due to arthritis-irAE	N/A	No	Yes
Duration of anti-IL-6R antibody therapy, months	12	15	3
Tumor response to CPI as of December 2016 (RECIST 1.1)	Complete remission	Progressive disease	Progressive disease

CPI, immune checkpoint inhibitor; UNL, upper normal limit; ANA, antinuclear antibody; CCP, anti-cyclin citrullinated antibody; RF, rheumatoid factor; Ro, anti-SSA antibody; dsDNA, anti-double-stranded DNA antibody; La, anti-SSB antibody; N/A, not applicable.

Methods: Three cases of a 78-year-old man with RP, a 74-year-old woman with RP associated with optic perineuritis and a 42-year-old woman with RP associated with uveitis were assessed by US before and after treatments.

Results: Ultrasonography (US) of the auricle before treatment showed low-echoic swollen auricular cartilage with increased power Doppler signals (PDS) in all cases. US findings corresponded to the above-mentioned biopsy findings. After treatment with prednisolone combined with methotrexate, the auricular swelling completely resolved in all cases. Then, US findings also showed dramatic reductions in swelling of cartilage with the decrease in PDS.

Conclusions: US imaging can be used to differentiate between inflammation, vascular lesions, and tumors in the ear pinna. RP could be differentiated from the damage of repeated trauma (i.e. rugby) with producing subperichondrial serous effusion. As in the present cases, US imaging of the external ear and auricular cartilage in RP also facilitates evaluation of auricular lesions and monitoring of disease activity, especially when we consider the treatment response and the timing of drug tapering.

Disclosure of Interest: None declared

DOI: 10.1136/annrhumdis-2017-eular.1769

FRI0604 SUCCESSFUL TREATMENT OF ARTHRITIS INDUCED BY CHECKPOINT INHIBITORS WITH ANTI-INTERLEUKIN-6 RECEPTOR ANTIBODY: A CASE SERIES

S.T. Kim¹, A. Diab², M. Uemura², S. Garcia³, P. Hwu², J. Tayar¹.

¹Rheumatology; ²Melanoma Medical Oncology, MD Anderson Cancer Center;

³Rheumatology, Baylor College of Medicine, Houston, United States

Background: Immune checkpoint inhibitors (CPIs) have significantly improved outcomes for patients with various cancers. However, CPIs are associated with immune-related adverse events (irAEs). Two to three percent of patients receiving a CPI develop arthritis. In general, severe irAEs are treated with a high-dose steroid and a tumor necrosis factor inhibitor (TNFi), usually infliximab; however, TNFi treatment is sometimes contraindicated and, furthermore, entails a theoretical concern of impairing antitumor immunity. Therefore, evidence showing the clinical benefits of non-TNFi biologics in the treatment of irAEs is of immediate interest.

Objectives: We described the effects of anti-IL-6R antibody for the treatment of arthritis-irAE.

Methods: Three patients receiving CPI developed arthritis and were treated with anti-IL-6R antibody. Patients were followed up to 15 months.

Results: All patients had metastatic melanoma (Table 1). No patient had a

Figure 1. Serial measurement of the patient's global assessment, tender joint count, and swollen joint count before (0 month) and after anti-IL-6R antibody treatment