arthritis (n=18, 16%), gynecological diseases (n=11, 10%) kidney stones (n=9, 8%) and others (n=34, 31%). As shown in Table, the frequency of patients with misdiagnosis, was significantly lower in Group 2 (66%) compared to Group 1 (84%).
A total of 59 patients (39%) received other long-term treatments, mainly monthly penicillin (n=28), prior to colchicine. There were 41 surgical interventions in 36 patients (24%), before the diagnosis of FMF, the most common being appendectomy in 31, gyneacological operations in 5, cholecystectomy in 3 and others in 2 occasions. It was noted that, the frequency of surgical operations was significantly decreased in Group 2 (12%) compared to Group 1 (27%) (Table).
The presence or absence of MEFV mutations was assessed in 69 patients (46%) before the diagnosis or after to reinforce the diagnosis. As expected, this was significantly more frequent in Group 2 (59%) compared to Group 1 (33%) (Table).
Seventy patients (46%) were diagnosed as FMF only after someone else in the family (n=44) or a friend (n=26) had a similar diagnosis. The frequency of these patients was similar when Group 1 and 2 were compared.

Table: Demographic and clinical characteristics of Group 1 (patients seen before 2000) and Group 2 (patients seen after 2000)

	Group 1, (n=70)	Group 2, (n=73)	P
Male/Female, n	22/48	19/54	Non significant
Current age, med [IQR] years	40[31-51.5]	32[24.5-38]	<0.001
Delay in diagnosis, med [IQR] years	10 [6-18]	4 [0-11]	<0.001
Misdiagnosed patients, n (%)	59 (84)	48 (66)	0.011
Surgery before diagnosis, n (%)	19(27)	9(12)	0.026
Assessment of MEFV mutations, n (%)	23 (33)	43 (59)	0.005
Diagnosed as FMF after someone else, n (%)	27(39)	38(52)	0.1

Conclusions: Although there is considerable decrease in delayed diagnosis of FMF, there is still significant amount of misdiagnoses after the year 2000, even in a geography where FMF is highly prevalent.
Table: Demographic and clinical characteristics of Group 1 (patients seen before 2000) and Group 2 (patients seen after 2000)
References:
[1] Tunca M et al; Turkish FMF Study Group.Familial Mediterranean fever (FMF) in Turkey: results of a nationwide multicenter study. Medicine (Baltimore). 2005 Jan;84(1):1–11.
Disclosure of Interest: None declared
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FRI0611 ASSESSMENT OF PANNICULITIS CLINICAL OUTCOMES: RISK FACTORS FOR RECURRENCE AND PREDICTORS OF SLOW REGRESSION OF INDURATIONS

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Background: Currently there's no clear understanding of the clinical course and outcomes of panniculitis with predominant involvement of subcutaneous adipose tissue (SAT), which is often associated with the involvement of locomotor system and viscera. Consistent elaboration of both is of paramount importance, as it may give a deeper insight into clinical and curative factors which may have impact on the disease prognosis.
Objectives: To assess clinical outcomes of panniculitis (risk factors for recurrence and predictors of slow regression of indurations).
Methods: 186 pts (172 females, 14 males) aged 43,5±14,5 years with different types of Pn, who were at the record of V. A. Nasonova Research Institute of Rheumatology during 2009–2015 yy. Disease duration varied from 1 week to 20 years. General clinical examination and lab tests (serum levels of α1-antitrypsin, amylase, lipase, ferritin, creatine phosphokinase (CPK), rheumatoid factor) (RF), ANF, anti-DNA, ANCA), bacteriological and serological tests, radiological examination (chest CT scan), ultrasound examinations of skin and subcutaneous fat (SF) around the nodules, tuberculin skin test and histopathology of nodular skin samples were performed at baseline. Patients were re-tested during the follow up (FUP) when necessary.
Results: Female middle-aged patients prevailed in the study group with the disease duration from 1 week to 25 years. *Saucer*-like depressions as a Pn - specific phenomenon was documented in 28,5% cases. The estimated risk of *saucer*-like depressions was higher in patients who manifested Pn at the age of >40 years (OR 4,9, 95% CI 2,3–10,4; $p<0,0001$); this subgroup also showed lesion proneness to confluence and forming of irregular shape conglomerates (OR 2,9, 95% CI 1,45–5,7; $p=0,002$), tendency to forming large size >3 cm lesions (OR 4,96, 95% CI 2,2–10,97; $p<0,0001$), and the disease lasting over 3 months (OR 24,9, 95% CI 9,2–67,4; $p<0,0001$). All pts were re-examined within 1–6 years. Median time to onset of nodules regression was 2 [1;6] months. Recurrences were documented in 66 (35,5%) pts during the follow up. Logistic regression was used for multivariate statistical analysis in order to identify the potential predictors of delayed (>3 months) nodules regression and recurrence risk factors. Used model identified the following predictors of delayed nodules regression (i.e., disease regression): age >40 years (OR 2,58; CI 1,02–6,5; $p=0,04$), and presence of saucer-like depressions (OR 5,05; CI 1,2–21,7; $p=0,03$). The sensitivity of used statistical model was 70%, specificity – 71%, positive predictive value - 74%. Disease duration >3 months (OR 4,7; CI 2,0–10,6; $p=0,0002$) was identified by our model as the predictor of recurrences with 60% sensitivity, 78% specificity, and 59% positive predictive value.
Conclusions: Pn tends to regress more slowly in pts aged over 40 and in pts having saucer-like depressions. Trend to recurrences is not so evident in Pn, although probability of recurrence increases in pts with longer disease duration at the time of initiation of therapy.
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FRI0612 IGG4-RELATED DISEASE IN EASTERN MEDITERRANEAN: CLINICAL FEATURES AND OUTCOMES OF A LARGE COHORT

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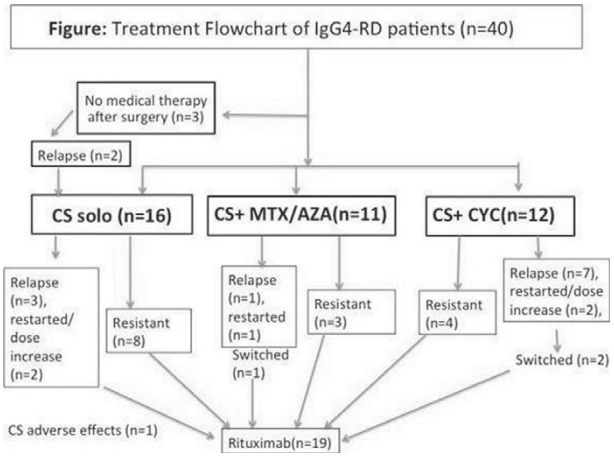
Background: Since majority of IgG4-related disease (IgG4-RD) patients in the literature are from Far East and United States, there is a lack of large series from other parts of the world.
Objectives: We aimed to identify the clinical characteristics and outcome of Turkish IgG4-RD patients from a tertiary center.
Methods: In step 1, clinical and histopathological features of patients having any disease under the clinical spectrum of IgG4-RD were retrospectively reviewed. For step 2, patients prospectively diagnosed are enrolled into the study. Patients fulfilling the "definite" diagnosis according to comprehensive diagnostic criteria were recruited (enrolled (n=52), excluded (n=47).
Results: Median age was 51.1 years and no gender predominance was observed (Male/female: 26/26). Retroperitoneal fibrosis is the most frequent presentation;

Table 1. Distribution of clinical findings and organ involvement (n=52)

Constitutional symptoms, n (%)	
Fatigue	31 (59.6)
Tiredness	16 (30.7)
Night sweats	15 (28.8)
Weight loss	14 (26.9)
Fever	13 (25)
Retroperitoneal fibrosis, n (%)	23 (44.2)
Lymphadenopathy, n (%)	20 (39.2)
Any cardiovascular involvement, n (%)	15 (28.8)
Periaortitis	12 (23.1)
Pericardium	5 (9.6)
Coronary periarteritis	4 (7.7)
Abdominal aort aneurysm	1 (1.9)
Orbital pseudotumor, n (%)	12 (23.1)
Orbitalmass/proptosis	6 (11.5)
Extraocular muscles	6 (11.5)
Pancreas, n (%)	12 (23.1)
Major salivary glands, n (%)	11 (21.2)
Lacrimal glands, n (%)	9 (17.3)
Mediastinal fibrosis, n (%)	6 (11.5)
Ear, nose, sinuses, n (%)	5 (9.6)
Lung fibrosis, n (%)	5 (9.6)
Skin , n (%)	4 (7.7)
Pleura, n (%)	4 (7.7)
Gall bladder and Biliary ducts, n (%)	4 (7.7)
Thyroid, n (%)	3 (5.8)
Liver, n (%)	3 (5.8)
Kidney (mass) , n (%)	3 (5.8)
Pachymeningitis, n (%)	2 (3.8)

Breast involvement: (n=1), tubulointerstitial nephritis (n=1).



others were lymphadenopathy, orbital pseudotumor, pancreas and salivary glands in decreasing order (Table). Twenty-four (46.1%) of patients had localized involvement. Corticosteroids were mainstay of treatment in 92.5% of patients, and in 57.5% with any immunosuppressive agents as first line treatment. Rituximab has been used for cases resistant to previous treatment or with relapses in 19 (47.5%) of patients. A complete response was achieved in 52.5% of patients and partial response (<50% of regression) in 40%. Two patients deceased due to IgG4-RD attributed problems and no malignancy was observed (median follow up: 18 months).
Conclusions: We observed similar features with previous European cohorts however no male predominance was seen. Even though conventional immunosuppressives were used in more than half of patients, treatment had switched to rituximab ~50% patients owing to resistance or relapses.
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FRI0613 H-FERRITIN AND PRO-INFLAMMATORY CYTOKINES ARE INCREASED IN THE BONE MARROW OF ADULT PATIENTS AFFECTED BY MACROPHAGE ACTIVATION SYNDROME

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Background: During macrophage activation syndrome (MAS), an inflammatory life-threatening syndrome, extremely high levels of serum ferritin may be observed [1]. Ferritin is an intracellular iron storage protein comprising 24 subunits that may be divided in heavy (H) subunits and light (L) subunits, based on their molecular weight [2]. The H-/L-subunits ratio may change, depending on the specific tissue and the physiologic status of the cell. In the normal condition, ferritin enriched in L subunits (L-ferritin) has been found in the liver and in the spleen, whereas the ferritin enriched in H subunits (H-ferritin), may be mainly observed in the heart and kidneys [2].
Objectives: We investigated the tissue expression of both H-and L-ferritin as well as the macrophage subsets expressing these molecules, in the inflammatory BM infiltrate of MAS patients. In addition, the co-expression of IL-1β, TNF, IFN-γ and H- or L ferritin, within the inflammatory cells, was assessed. Finally, we explored if the imbalance between H-ferritin and L-ferritin as well as the number of ferritin positive cells may be considered helpful bio-markers to assess the severity of these patients.
Methods: We analysed the bone marrow (BM) biopsies, by immunofluorescence of 10 adult MAS patients affected by rheumatic disease to assess the presence of: i. both H- and L-ferritin; ii. the number of CD68+/H-ferritin+ and CD68+/L-ferritin+; iii. the tissue pro-inflammatory cytokines, IL-1β, TNF, IFN-γ; and we correlated these data with clinical and laboratory data. Furthermore, the presence of ferritins was assessed in the sera of the same patients by western blot analysis.
Results: We observed an increased tissue expression of H-ferritin and of pro-inflammatory cytokines (IL-1β, TNF, IFN-γ). Western blot analysis, in the sera, of H-ferritin mirrored data on the tissue. Furthermore, an increased number of CD68+/H-ferritin+ cells and an infiltrate of cells co-expressing H-ferritin and IL-12, suggesting an infiltrate of M1 macrophages, were observed.
Tissue H-ferritin levels correlated with the decreased counts of WBC (p=0.01) and PLT (p=0.0001); with the increased values of serum ferritin (p=0.012) and C-reactive protein (CRP) (p=0.0058); and with the tissue expression of IL-1β (p=0.006). The number of the CD68+/H-ferritin+ cells correlated with the decreased counts of WBC (p=0.03) and PLT (p=0.0007), and with the increased serum ferritin levels (p=0.0088) and CRP (p=0.049). The analyses concerning tissues L-ferritin as well as the number of CD68+/L-ferritin+ cells and the same parameters failed to show any significant result.
Conclusions: We observed an increased tissue expression of H-ferritin associated with an increased expression of IL-1β. Interestingly, in the BM inflammatory infiltrate an increased number of CD68+/H-ferritin+ cells was shown. Of note, tissue expression of H-ferritin as well as the number of CD68+/H-ferritin+ significantly were associated with the hematological involvement of the disease, suggesting possible bio-markers to assess the severity of these patients.
References:
[1] Ramos-Casals M, et al. Lancet. 2014;383:1503–16.
[2] Rosário C, et al. BMC Med. 2013;11:185.
Disclosure of Interest: None declared
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FRI0614 FREQUENCY OF ORGAN MANIFESTATIONS IN CHRONIC SARCOIDOSIS

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Background: Chronic sarcoidosis is a systemic disease of unknown etiology, characterized by the histological finding of granulomas in involved organ systems. The most often affected organ is the lung with approximately 90–95%. Systematic data of organ manifestations other than the lung are scarce and show a wide range from 1–2% up to 50% depending on the series.
Methods: We analyzed data of newly diagnosed chronic sarcoidosis in 3 tertiary hospitals. We analyzed data on organ manifestations (OM), type of OM and laboratory findings. The certainty of OM was classified as grade 0 (not investigated), grade 1 (no sign of OM), grade 2 (clinical sign of OM), grade 3 (signs of OM in laboratory findings or imaging) and grade 4 (histological proven OM), respectively.
Results: We included 151 patients with biopsy-proven chronic sarcoidosis. Mean age was 50.8±15 years with a male predominance (87 [57.2%] vs. 65 [42.8%] patients).
Except for 3 patients, all demonstrated pulmonary involvement. The predominant type of lung involvement was type I (mediastinal lymph node enlargement) in 54.2% and type II (mediastinal lymph node enlargement and interstitial involvement) in 27.7%.
96.5% of patients were investigated for an affection of the kidneys, 97.3% for hepatic, 92.7% for skin involvement, 68.8% for ocular manifestations, 67.5% for ear, nose, throat (ENT) manifestations and 92% for cardiac manifestations, respectively.
Grade 3 (imaging/laboratory) and grade 4 (histology) findings were seen in the kidneys in 7.6/22.8%, in the liver in 13.3/11.9%, in the heart in 10.6/0.7%, in the eyes in 6.6/
Conclusions: OM in chronic sarcoidosis are more frequent than suggested in the current literature, especially renal and hepatic. About 20% of patients with chronic sarcoidosis suffered from moderate to severe CKD due to sarcoidosis, which is a major organ complication contributing to overall morbidity.
We recommend a systematic screening for OM in all patients with chronic sarcoidosis as it is performed in other systemic rheumatic disease.
Disclosure of Interest: None declared
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FRI0615 SARCOIDOSIS AND CANCER: DIFFERENT PATTERNS OF ASSOCIATION IN A MULTICENTER COHORT FROM SOUTHERN EUROPE

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Objectives: To evaluate the temporal association between the diagnosis of neoplasia and sarcoidosis in a large cohort of Spanish patients with sarcoidosis.
Methods: In January 2016, the Autoimmune Diseases Study Group (GEAS-SEMI) created a national registry (SARCOGEAS) of patients with sarcoidosis. Sarcoidosis was diagnosed with the criteria proposed by the ATS/ERS/WASOG 1999 statement, and extrathoracic disease by the 2014 WASOG instrument. Diagnosis of neoplasia was recorded before and after the diagnosis of sarcoidosis.
Results: The cohort included 1082 patients (82% biopsy-proven, 618 women, mean age 47yrs). Association with neoplasia was detected in 135 (13%) patients who developed 140 neoplasms (110 solid and 30 hematological neoplasia). The neoplasia more frequently reported were breast (n=18), lymphoma (n=16), non-melanoma skin (n=15) and colon (n=15). Association with neoplasia was more frequent in patients born in Spain (97% vs 86%, p<0.001, OR 4.06), older patients (55 vs 46yrs, p<0.001, OR 1.03) and those with bone marrow involvement (14% vs 4%, p<0.001, OR 3.64). Patients in whom cancer preceded the diagnosis of sarcoidosis had a higher frequency of sarcoidosis diagnosed incidentally (20% vs 4%, p=0.011) and a lower frequency of ocular sarcoidosis (3% vs 16%, p=0.016). Patients with associated hematological neoplasia had a higher frequency of ENT (13% vs 1%, p=0.009) and bone marrow (33% vs 9%, p=0.002) involvements in comparison with patients with associated solid neoplasia.
Conclusions: Association between sarcoidosis and cancer was found in 13% of patients (80% solid and 20% hematologic malignancies). Elderly patients and those born in Spain were at high risk of having associated cancer. Asymptomatic