

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, gynecological 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.

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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  $\alpha$ 1-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 (%)	
Lymphadenopathy, n (%)	23 (44.2)
Any cardiovascular involvement, n (%)	20 (39.2)
Periaortitis	15 (28.8)
Pericardium	12 (23.1)
Coronary periarthritis	5 (9.6)
Abdominal aort aneurysm	4 (7.7)
Orbital pseudotumor, n (%)	1 (1.9)
Orbital mass/proptosis	12 (23.1)
Extraocular muscles	6 (11.5)
Pancreas, n (%)	6 (11.5)
Major salivary glands, n (%)	12 (23.1)
Lacrimal glands, n (%)	11 (21.2)
Mediastinal fibrosis, n (%)	9 (17.3)
Ear, nose, sinuses, n (%)	6 (11.5)
Lung fibrosis, n (%)	5 (9.6)
Skin, n (%)	5 (9.6)
Pleura, n (%)	4 (7.7)
Gall bladder and Biliary ducts, n (%)	4 (7.7)
Thyroid, n (%)	4 (7.7)
Liver, n (%)	3 (5.8)
Kidney (mass), n (%)	3 (5.8)
Pachymeningitis, n (%)	2 (3.8)

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

