Table 1. Demographic and clinical characteristics of the patients

	MEFV gene mutations	Treatment cycles (months)	Attacks per year before TCZ	Attacks per year after TCZ
44/Female	M694V homozygote	96	24	5
49/Male	M694V homozygote	48	30	0
59/Male	M694V/N heterozygote	8	12	1
79/Male	M694V/M680I heterozygote	67	12	0
40/Female	M694V homozygote	2	14	1
38/Male	M694V homozygote	3	48	3
44/Male	M694V homozygote	18	30	10
49/Male	M694V homozygote	3	36	0
34/Male	M694V homozygote	23	24	0
39/Male	MEFV/- heterozygote	33	20	0
28/Male	M694V homozygote	19	18	0
24/Male	M694V homozygote	12	30	2
27/Male	M694V homozygote	3	36	36
27/Male	M694V/M680I heterozygote	12	12	0
50/Male	M694V homozygote	12	6	0

MEFV:Mediterranean Fever, TCZ:Tocilizumab

Conclusion: Tocilizumab is found to be effective on improvement of amyloidosis. In the current analysis, we found that TCZ also decreases the frequency of recurrent attacks in patients with FMF. Besides, TCZ is well tolerated among the patients. Further and prospective studies with larger sample are needed to support these results.

REFERENCES:

- [1] Yilmaz S, Cinar M, Simsek I, Erdem H, Pay S. Tocilizumab in the treatment of patients with AA amyloidosis secondary to familial Mediterranean fever. *Rheumatology* 2015;54:564-565

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FRI0587 IGG4-RELATED DISEASE IN ITALY: RESULTS FROM A MONOCENTRIC COHORT OF 150 PATIENTS (2013–2018)

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Background: IgG4-related disease (IgG4-RD) is a recently recognized fibro-inflammatory disease characterized by fibrotic lesions and elevated IgG4 serum level. Epidemiological and clinical data in our country have not been precisely assessed yet

Objectives: The aim of this work is to present the data from the largest monocentric Italian IgG4-RD cohort.

Methods: We conducted a retrospective analysis searching for IgG4-RD patients referred to our tertiary care centre between 2013 and 2018. Diagnosis were made according to the "Comprehensive Diagnostic Criteria 2011", and patients were clustered on the basis of a definite, probable and possible, diagnosis. Each patient underwent clinical, serological, and radiological evaluation according to the organ involvement. IgG4-RD activity was measured with IgG4-RD Responder Index (IgG4-RD RI). All the data are reported in median and interquartile range.

Results: 150 patients were included in this work (111 male, 39 female, M:F ratio 2.8:1). 75 cases received a definite IgG4-RD diagnosis, while 6 patients and 49 patients were diagnosed as *probable* and *possible* IgG4-RD, respectively. Median age at diagnosis was 60 (SD 13) years old. The most involved organ was the pancreas (66 cases; 44%), followed by lymph nodes (26 cases; 17%), biliary tree (25 cases; 17%), salivary glands (21 cases; 14%), aorta and retroperitoneum (12 cases; 8%), meninges (9 cases; 6%), nasal sinuses and nasal septum (5 cases; 3%). 18 patients (12%) had a positive history of malignancies that occurred ten years earlier or after IgG4-RD diagnosis. At baseline, IgG4 level was

elevated in 92 patients (61%), while eosinophil count and IgE level were elevated in 35 (23%) and 52 (35%) patients, respectively. Circulating plasmablast were measured in 88 and elevated in 66 (1859 cell/mL, 600-3950). Median IgG4-RD RI at diagnosis was 6 (6-9). 55 patients were treated with glucocorticoids alone, whereas immunomodulator (Mycophenolate mofetil, azathioprine, cyclophosphamide) were associated in 31 cases. Rituximab was administered in 32 patients. 15 patients underwent surgical excision of the fibrotic lesion. 15 patients were candidate to a watchful waiting strategy, due to low disease burden.

Conclusion: Our study reports clinical and epidemiological data from the largest monocentric cohort of patients with IgG4-RD in Italy. Our data reflect those reported in other international cohorts of IgG4-RD patients.

REFERENCES:

- [1] Della Torre E, Lanzillotta M, Doglioni C. Immunology of IgG4-related disease. *Clin exp Imm*.2015

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FRI0588 THE SCALE OF ACTIVITY IN IDIOPATHIC LOBULAR PANNICULITIS

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Background: Idiopathic lobular panniculitis (ILP) refers to chronic systemic connective tissue lesions (M 35.6). The disease develops with typical clinical symptoms and is characterized by alternating periods of stable condition (remission) and active manifestations (exacerbations). Assessment of the activity and identification of the disadvantageous predictors is the main task in monitoring patients with ILP, however, the "gold standard" has not been developed yet.

Objectives: to create a rating scale for the inflammatory process activity in ILP on the basis of clinical and laboratory parameters

Methods: We examined 67 patients (9 men and 58 women) with the ILP diagnosis verified in the V.A. Nasonova Research Institute of Rheumatology in 2007-2017. The age of patients ranged from 20 to 76 years, the average duration of the disease was 78.91[48;50] months. At the time of treatment at the institute only 16 patients (23,88%) were diagnosed with ILP. All the patients underwent comprehensive clinical examination and laboratory and instrumental examination of biochemical and immunological parameters, radiography or computed tomography (CT) of the chest organs, as well as pathomorphological examination of skin and subcutaneous fat biopsies from the node area.

Results: Analysis of clinical manifestations allowed to identify four forms of ILP: nodular (30 patients), plaque (10), infiltrative (15) and mesenteric (12), which had specific clinical and laboratory features. Based on the obtained data we developed a scale of the ILP activity (SA), which includes a description of the state of the 7 organ systems. The maximum score for individual systems is from 1 to 3 points depending on the number of estimated parameters. The total maximum possible score is 42 points. The score includes all types of damage since the onset of the disease (caused directly by ILP or developed as a result of therapy), while taking into account only signs that persist for 6 months. Score <5 shows inactive disease, 5-10 - low disease activity; 11-20 - moderate activity and >20 - high activity of the disease. Thus, low disease activity is characterized by the predominance in the clinical picture of limited moderately painful indurations in the absence of any significant changes in laboratory tests. At the moderate disease activity, changes in various localization with a predominance of proliferative disorders (common painful compaction of the 1st and/or 3rd stage, moderate area of damage to the trunk and limbs, subfebrile temperature, cognitive impairment, mesenteric disorders, etc.) are revealed against the background of less pronounced laboratory tests. ILP with high activity is characterized by the presence of fever and other common signs of the disease, the predominance of the 2nd stage with a pronounced VAS pain intensity of the induration, the common nature of the defeat of the pancreas, involvement of the lungs, heart and gastrointestinal tract in the pathological process.

Conclusion: In fact, SA is an analysis of the organ damage, assessment of the symptoms severity, and it takes into account the development of exacerbations with the involvement of new organs in the process of the disease. The proposed ILP SA is of practical importance. Further