Methods: Data of IgG4-RD patients treated with RTX and followed-up for >18 months were reviewed. Patients were treated with two 1 g RTX doses 15 days apart (standard dose, SD) and then divided into: patients receiving RTX in case of relapse (Group 1) and patients receiving maintenance RTX treatment 6 monthly (Group 2). Patients in Group 2 received a first maintenance treatment RTX SD and were then further divided into patients receiving RTX SD (Group 2a) or single 1 g RTX dose (Group 2b).

Results: 14 IgG4-RD patients were included. Median number of organs involved was 3 (1-5). Baseline median physician global assessment (PGA) was 70 (50-90), IgG4-RD responder index (IgG4-RD RI) 12 (6-12). Median eosinophil count, ESR, CRP, and serum IgG4 level were 100 cells/ μ L (50-1,800), 19 mm/h (6-64), 3.7 mg/L (0.1-36) and 3.17 mg/L (470-21,800) respectively. Indications for RTX were: relapse in 10/14 (71%), partial response in 3/14 (21%) and first-line treatment in 1 (7%) patient. CS were stopped in 5 patients and reduced by month 3 in remaining patients (27.14 $\pm$ 13.06 vs 6.43 $\pm$ 2.45 mg daily, $p=0.012$). 3 patients were on concomitant methotrexate (MTX) therapy at RTX induction, MTX was stopped in all patients at month 3. At month 6 PGA, IgG4-RD RI, and serum IgG4 decreased to a median value of 10 (0-50), 2 (0-6), and 822 mg/L (380-7,313), respectively ($p<0.001$ for all comparisons). Median eosinophil count, CRP and ESR levels did not change significantly. 8 (57.1%), 5 (35.7%) and 1 patient (7%) were on disease remission (DR), complete remission (CR), and partial remission (PR), respectively. Median number of RTX courses was 2 (1-2) in Group 1, 3 (3-7) in Group 2a, and 4 (3-5) in Group 2b. Organ involvement, PGA, IgG4-RD RI, ESR, CRP, and serum IgG4 levels were not significantly different between the 2 Groups at baseline. FRR at 18 months in Group 1 was significantly lower than that in Group 2 (29% vs 100%, $p=0.006$). 5 (71%) patients in Group 1 relapsed on average 9 (7-12) months after RTX induction. 2 (14%) patients had hypersensitivity reactions during RTX induction. 6 (43%) patients, 3 in Group 1 and 3 in Group 2, experienced low-grade infections.

Conclusion: RTX is a safe and effective treatment in IgG4-RD. Both SD and single 1 g RTX dose seem effective in maintenance.

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FRI0585 EFFICACY OF CANAKINUMAB AS FIRST-LINE BIOLOGIC AGENT IN ADULT-ONSET STILL'S DISEASE

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Background: Adult-onset Still's disease (AOSD) is a rare auto-inflammatory condition characterized by fever, arthritis, skin rash, and multi-organ inflammation. The pathogenesis of AOSD is centrally mediated by the pro-inflammatory cytokine interleukin (IL)-1 β ; anakinra, a recombinant inhibitor of the IL-1 β receptor, has thereby become the cornerstone of biologic therapy for AOSD. More recently, a new agent blocking IL-1 β has become available, that is, the monoclonal antibody canakinumab.

Objectives: So far, canakinumab has been used in the treatment of AOSD following failure of multiple lines of biologic therapy, including anakinra. Here, we describe the use of canakinumab as first-line biologic agent in 6 patients with AOSD, and report the highly promising results of this treatment approach.

Methods: Six patients with severe, DMARD-refractory AOSD received canakinumab at the approved dose of 4 mg/kg once every 4 weeks following failure of conventional treatment with corticosteroids and methotrexate. Patient characteristics and response to therapy were recorded.

Results: In all patients, treatment with canakinumab led to striking and rapid clinical responses, within days of initiation. Fever and skin rash disappeared first and did not recur, followed by progressive and sustained

improvement in arthritis. Inflammatory organ involvement also responded to treatment: for example, we observed resolution of pericardial inflammation in two patients, as revealed by cardiac ultrasound; another patient had hepatosplenomegaly, which also reverted to normal upon treatment. A striking reduction in serum pro-inflammatory markers C-reactive protein, erythrocyte sedimentation rate, and ferritin mirrored the efficacy on clinical manifestations. These effects amounted to a significant decrease in the modified Pouchot score for measuring disease severity (average score before canakinumab treatment: 6; after: 1.25; statistical significance of difference evaluated with Mann-Whitney: $p=0.02$), and allowed for tapering or discontinuation of concomitant therapy: specifically, corticosteroid treatment was completely discontinued in two patients and substantially reduced in two patients, whereas methotrexate was stopped in one patient.

Conclusion: In this study, first-line biologic therapy of AOSD with canakinumab resulted in rapid and marked efficacy, ultimately leading to full clinical remissions in all patients, and allowing for robust steroid-sparing effects. So far, canakinumab in AOSD has mostly been used as a last line of treatment following failure of multiple other agents, including anakinra. Early treatment, as described in this study, is nevertheless advisable and may reduce chances of chronic disease and permanent damage.

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FRI0586 THE EFFECT OF TOCILIZUMAB IN THE PREVENTION OF FAMILIAL MEDITERRANEAN FEVER ATTACKS

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Background: Familial Mediterranean Fever (FMF) is the most frequent hereditary autoinflammatory disease in the worldwide. Colchicine is the mainstay of the treatment that is used for both preventing the attacks and developing amyloidosis. In case of colchicine resistance, which constitutes almost 5% of all patients, there are some alternatives such as anti-TNF agents, IL-6 and IL-1 antagonists. These agents have also been used in patients with established amyloidosis secondary to FMF. Tocilizumab (TCZ), an IL-6 receptor antagonist, although is effective in the treatment of amyloidosis, little known about its effectiveness in the preventing of febrile attacks.

Objectives: In this study, we aimed to find out whether TCZ therapy decrease the frequency of recurrent attacks of FMF or not.

Methods: This retrospective cross-sectional study designed in a tertiary rheumatology clinic between the years 2009 and 2019. A total of 15 patients were given intravenous TCZ at a dose of 8 mg/kg/month, to improve amyloidosis associated with FMF. Data of demographic and clinical characteristics of patients were obtained retrospectively from patient files.

Results: Three female and 12 male patients received TCZ for amyloidosis treatment. The mean age of patients was 42.07 $\pm$ 14.37 years and disease duration was 25.73 $\pm$ 10.86 years. All of the patients had biopsy results providing amyloidosis. According to international severity scoring system for FMF, all of the patients had severe disease. Demographic and clinical data were shown in Table 1. The frequency of attacks was evaluated during TCZ treatment, and it was reported that 1 patient had no response, 6 patients had decreased attack frequency and 8 patients had no attacks. Serious adverse events were seen in none of the patients.

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.

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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.

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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