a trend in favor of ADA, except for Sarcoidosis subgroup in the VISUAL II trial (Figure). Overall safety for both trials has been previously reported ^{1,2}.

Conclusions: These exploratory analyses from the VISUAL I and VISUAL II trials show significantly higher efficacy in ADA-treated patients over PBO in "idiopathic/other" diagnoses of patients with both active and inactive non-infectious uveitis. Furthermore, across different uveitis etiologies, these analyses suggest that ADA-treated patients had a prolonged time to treatment failure compared to PBO.

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FRI0619 COMPARING CANAKIMUMAB AND ANAKINRA IN YOUNG GREEK WOMEN WITH RESISTANT RECURRING PERICARDITIS

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Background: Resistant – recurring pericarditis (RRP) is often associated with autoimmune syndromes. Conventional therapy with glucocorticoids and colchicine is often enough unsuccessful, thus nowadays the use of anakinra in such cases is becoming common every – day practice.

Objectives: Our goal was to determine whether RRP was associated with gene mutations relevant with autoinflammatory diseases and then compare the effectiveness of canakinumab and anakinra.

Methods: We studied 18 patients with RRP (all women, from 14 to 36 years old) determining the existence of mutation from the Infevers database (25 genes, from MEFV and TNFRSF1A to MVK and SLC 29A3, all related with autoinflammatory diseases). All the patients were proven positive to one of the above mutations (all heterozygous) and then treatment with anakinra (n=9, 100mg sc once daily) and canakinumab (n=9, 150mg sc q4wk) was administered.

Results: During the study (3 years), all anakinra patients stopped receiving per os glucocorticoids and were free of any new pericarditis episodes. 3 of them (33%) stopped any treatment after a year of anakinra therapy and none was presented with any skin reactions.

On the contrary, all canakinumab patients were not able to free themselves of the glucocorticoid treatment, since at least one new episode of pericarditis to each patient was recorded. Two of them were free of pericarditis the last year of the study (though being treated with 2,5mg prednisolone per os daily).

Statistically speaking, the use of IL-1R antagonist in recurrent – resistant pericarditis associated with autoinflammatory mutations was proven much more successful (p<0,01) than the use of the IL-1b Mab.

Conclusions: Though the sample of patients (n=18) was too small in order to set us able to reach any safe conclusions, it is quite possible that anakinra may be one effective solution that can prevent the long term administration of corticoids to patients with recurrent pericarditis. Canakinumab was not recorded with similar positive results.

Disclosure of Interest: None declared

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FRI0620 MIR-204-3P INHIBITS THE PRODUCTION OF TLR4-RELATED CYTOKINES IN FAMILIAL MEDITERRANEAN FEVER BY TARGETING THE PIK3 SIGNALING PATHWAY

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Background: MicroRNAs (miRNAs) are endogenous small RNAs and post-transcriptionally regulate gene expression by pairing with target. There has been

emerging evidence showing the association of aberrantly expressed circulating miRNAs in the serum with the pathogenesis or progression of diseases including cancer and autoimmune disease. Although a number of circulating miRNAs associated with inflammation have been identified, the roles of them in patients with FMF and the underlying mechanism remain to be elucidated.

Objectives: The aim of this study was to identify a serum miRNAs profile and potential biomarkers in FMF and clarify their gene targets for understanding the pathogenesis of autoinflammatory diseases.

Methods: We performed miRNA microarray in the serum from FMF in attack and in remission. We subsequently examined the expression of miRNAs that varied before and after the attack in macrophages derived from THP-1 cells stimulated with toll-like receptor (TLR) ligands. Macrophages derived from THP-1 cells transfected with pre-miRNA and anti-miRNA were stimulated with TLR ligands for 24 hours. We collected the supernatants for the quantification of inflammatory cytokine production. To identify the target genes, we overexpressed its miRNA and performed Agilent expression microarray. Transfection with reporter construct and pre-miRNA and anti-miRNA was performed to confirm suppression of target mRNA.

Results: We found that miR-204–3p was greatly decreased in the serum from FMF patients in attack. In vitro study, the expression of miR-204–3p was suppressed by LPS stimulation in macrophages derived from THP-1 cells. Inhibition of miR-204–3p significantly induced the production of TLR4-related cytokines whereas overexpression of miR-204–3p inhibited their production. Bioinformatic analysis showed that miR-204–3p is predicted to target genes implicated in TLR pathway through regulation of PIK3 signaling. Reporter assay revealed that miR-204–3p directly suppressed the luciferase activity of 3'UTR of PIK3CG reporter construct.

Conclusions: These data suggest that serum miR-204–3p has a potential as a useful biomarker among patients with FMF and that miR-204–3p plays a critical role as a suppressor to regulate the production of TLR4 related cytokines by targeting PIK3 signaling pathway.

Disclosure of Interest: None declared

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FRI0621 PREVALENCE OF DIFFERENT ORBITAL ANATOMIC STRUCTURES AFFECTION IN IGG4-RELATED OPHTHALMIC DISEASE: SINGLE CENTER EXPERIENCE

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Background: According to 2009 Japanese nationwide survey of IgG4-related disease (IgG4-RD), orbit is the leading site of affection [1]. However diagnostic criteria and nomenclature of IgG4-RD were developed a few years later [2,3].

Objectives: To evaluate the peculiarities of clinical, laboratory, histological and immunohistochemical presentation of IgG4-related ophthalmic disease (IgG4-RD).

Methods: During 2004 – 2016, 82 patients were diagnosed with IgG4-RD 53 of whom had IgG4-ROD (men – 17, women - 36). The diagnoses of IgG4-RD and IgG4-ROD were established using comprehensive diagnostic criteria [2,3]. In all patients full clinical, ophthalmological, dental and serological (rheumatoid factor, C-reactive protein, IgG, IgG4, IgM, IgA, ANA, anti-Ro/La, C3/C4 complement) examination was carried. In all cases diagnosis was verified pathomorphologically with immunohistochemical staining (anti-CD 138, CD 68, IgG, IgG4, κ-chain, λ-chain), but only in 43 patients the diagnosis of IgG4-RD was established on orbital tissues biopsy. Some patients at baseline were tested on B-cell clonality

in tissue (on frozen tissue section or paraffin embedded) by PCR analysis of immunoglobulin V-D-J genes heavy chain rearrangements (FR1, FR2, FR3); monoclonal secretion was tested in serum protein electrophoresis.

Results: 40 patients with IgG4-ROD had orbital lesions at the onset of the disease and 13 developed them after 2 - 9 years from the onset. The frequency of different orbital anatomic structures involvement see in table 1. 15 (28%) patients had isolated orbital lesions while 38 (72%) had systemic IgG4-RD with simultaneous involvement of 2 - 7 other organs. 6 (14%) patients had unilateral orbital lesions. 23 (53%) patients with IgG4-ROD had associated IgG4-related sialoadenitis and nasal lesions. Most common pathomorphological features of IgG4-ROD see in table 2. The most common laboratory features of IgG4-ROD were elevation of: total serum IgG (44%), serum IgG4 (88%), serum IgE (61%) and monoclonal serum secretion (23%).

Table 1. Prevalence of different orbital anatomic structures affection in IgG4-ROD (n=43)

Anatomic structure	N, %
Lacrimal gland	93
Extraocular muscles	85,5
Fibroinflammatory lesions of eyelids	32,5
Retrolbulbar infiltration	32,5
Optic nerve thickening	32,5
Trigeminal involvement	21
Nasolacrimal duct	4,5

Table 2. Pathomorphological features in IgG4-ROD (n=43)

Pathomorphological feature	N, %
Lymphoplasmatic infiltrate	93
Dense fibrosis	93
Lymphoid follicle formation	69
Tissue eosinophilia	32,5
Obliterative phlebitis	14
Vasculitis	7
IgG4/IgG > 40%	90,5
MALT-lymphoma of lacrimal gland	2,3
AL-amyloidosis	2,3

Conclusions: In our cohort of IgG4-RD patients orbit is the leading site of the disease (64.5% of patients) and the disease onset (75% of patients). The majority of patients have bilateral orbital lesions (89%) and systemic course of the disease with other organ involvement (72%). Most often affected orbital anatomic structures are lacrimal glands, extraocular muscles, retrolbulbar infiltration with optic nerve thickening and fibroinflammatory lesions of eyelids. MALT-lymphoma of lacrimal gland and local AL-amyloidosis can complicate the long history of IgG4-ROD. Monoclonal serum secretion and B-cell clonality in the tissue in 23% of patients can potentially act as a background for lymphoma formation.

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FRI0622 A RETROSPECTIVE COHORT STUDY ABOUT THE CLINICAL CHARACTERISTICS OF IGG4-RELATED DISEASES

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Background: Immunoglobulin (Ig) G4-related disease (IgG4-RD) is an increasingly recognized systemic autoimmune disease in the past decade, and encompasses a wide spectrum of disorders previously considered to be unrelated, but with common pathologic, serologic, and clinical features. There have been several hundred literatures on IgG4-RD published in the past few years, however, we still cannot well characterize this complicated disease, including clinical features, treatments as well as mechanisms, which promotes us to further describe this disease.

Objectives: To describe the clinical presentations, laboratory features, imaging manifestations, treatments and evolutions in a cohort of 34 Chinese patients with immunoglobulin G-related diseases (IgG4-RD).

Methods: A retrospective study was performed in Wuhan Tongji Hospital. Electronically stored records of clinical, laboratory, imaging, and histological features were reviewed. The diagnoses were made according to comprehensive diagnostic criteria for IgG4-RD. Disease activity was assessed by IgG4-RD Responder Index (RI). This study was approved by the Ethical Committee of Tongji Hospital (TJ-C20150603). All patients were contacted, and consents were obtained from all the participants.

Results: We found that among the whole 34 patients enrolled in this study (median

age=51 years, male/female ratio=1.4/1), 65.2% tested patients had a declined HDL level, 82.3% tested patients had an elevated IgG4 level, and lymph node was the most frequently involved organ. 18F-FDG PET/CT had high sensitivity in providing information about the organ distribution of IgG4-RD. The IgG4 level and IgG4-RD RI in the multi-organs affected patients were significantly higher than that in the localized ones. Most patients were prescribed with glucocorticoids (GCs) alone or combined with disease modifying antirheumatic drugs (DMARDs), the majorities responded well, while some relapsed. Patients given rituximab (RTX) obtained good responses.

Table 1. The overall prognosis of 19 patients who were followed up

Prognosis	GCs	GCs + DMARDs	Surgery + GCs	Surgery + DMARDs	Antifungal therapy	MMF	Total, n
Remission, n	5	1	2	0	0	1	9
Recurrence, n	3	4	0	0	0	0	7
Worsen/Persistence, n	0	1	0	1	1	0	3
Total, n	8	6	2	1	1	1	19

Abbreviations: GCs, glucocorticoids; DMARDs, disease-modifying anti-rheumatic drugs; MMF, mycophenolate mofetil.

Fig 1: Analysis of IgG4, IgG4/IgG(%), IgG4-RD RI, and ESR levels between patients of single organ involvement and multiple organs involvement.

Conclusions: It was concluded that 18F-FDG PET/CT can be an effective tool to detect involved lesions of IgG4-RD. The addition of DMARDs to the initial GCs therapy in the remission induction stage may not bring much benefit. The most appropriate dosage schedule of RTX needs to be further investigated. Patients with younger age, longer duration of disease, higher IgG4 level and IgG4/IgG ratio may more commonly experience recurrence.

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Diagnosics and imaging procedures

FRI0623 [18F]FLUORIDE PET-CT IMAGING OF BONE FORMATION IN ANKYLOSING SPONDYLITIS BEFORE AND AFTER 12 WEEKS OF ANTI-TNF TREATMENT

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Background: Excessive bone formation is an important hallmark of ankylosing spondylitis (AS). Recently, it has been demonstrated that axial bony lesions in AS patients can be visualized using [¹⁸F]fluoride PET-CT. This technique may also provide opportunities to monitor changes in bone formation in axial AS lesions during therapy.

Objectives: To assess (1) changes in [¹⁸F]fluoride uptake in axial lesions of AS patients before and 12 weeks after treatment with TNF-inhibitors (TNFi) and (2) whether [¹⁸F]fluoride uptake on PET is related to focal bone formation in spine biopsies.