HLA B7 and HLA B35 were the most frequent genotypes (figure 1). Sub group analysis showed that 87.5% of patients with a component of posterior uveitis were HLA B7 or HLA B35 positive. It was noted none of 5 cases of HLA B35 +ve uveitis needed second line DMARD initiation after a tapering course of systemic steroids. There was no other significant difference in the clinical or treatment parameters noted in the subgroup analysis between the HLA genotypes.

Conclusion: Autoimmune uveitis not associated with HLA B27 tends to have older age of onset, female predominance, more common involvement of posterior uvea and association of vasculitis than HLAB27 autoimmune uveitis. Among non HLA B27 uveitis, HLA B07 and HLA B35 are the most common serotypes in our Asian south Indian group. Therapeutic stratification for a delayed initiation of second line DMARD therapy needs to be studied further particularly in HLA B35 +ve uveitis.

References:

- [1] Ninan F, Mathew J, Philip SS, John D, Danda D, John SS. Uveitis of spondyloarthritis in Indian subcontinent: a cross sectional study. *Int J Adv Med* 2017;4:1441-6

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SAT0539 MUCKLE-WELLS SYNDROME IN RHEUMATOLOGY PRACTICE, FAMILY CASES: FEDERAL CENTER EXPERIENCE.

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Background: Muckle-Wells syndrome (MWS) is a monogenic autoinflammatory disease caused by a NLRP3 gene mutation. It is the most common variant of cryopyrin-associated periodic syndromes (CAPS) and can be observed in rheumatology practice. It manifests itself in fever, urticaria-like rash, arthralgias/arthritis, conjunctivitis/uveitis, sensorineural hearing loss, acute-phase markers (ESR, CRP). The disease's onset usually takes place in infancy. There are examples of family cases. Targeted therapy: interleukin-1 inhibitors (anakinra, canakinumab).

Objectives: to provide characteristics of MWS patients, family cases in the rheumatology practice of the Federal Rheumatology Center in Russia.

Methods: in a 10-year period (2009 to 2019), MWS was diagnosed in 42 outpatient and inpatient patients, among them were 24 children, 18 adults, and 9 family cases. All of them underwent a standard rheumatology examination, including a ECR, CRP, ophthalmologist examination, and an audiogram. A molecular genetic test of the NLRP3 gene was carried out for all patients, the diagnosis was confirmed in all of them.

Results: Out of 18 adult patients aged between 19 and 59 years, women were prevalent (16 to 2), the onset age was 0 to 53 years, in 88.9% cases the onset took place before a patient was 18 years old. When diagnosed, the disease duration varied from 6 to 46 years. Most patients demonstrated fever, urticaria-like rash, arthralgias/arthritis, which were observed in 16 patients (88.9%), conjunctivitis were observed in 15 patients (83.3%), sensorineural hearing loss – in 8 patients (44.4%), abdominal pain, nausea, vomiting – in 4 patients, headache, dizziness – in 6 patients. There also were rare manifestations, such as: recurrent oral ulcers (8), genital ulcers (3), erythema nodosum (3), sore throat and raids on the tonsils (PFAPA-like phenotype) was observed in 2 patients. In 3 patients manifestations were triggered by cold temperature. All patients had an increased ESR and C-reactive protein concentration. Eight family cases of MWS were identified (in total 26 family members aged between 2.5 and 62 years) with a number of affected in one family ranging from 2 to 6 people of different age (8 children, 18 adults, out of which 20 were female, and 6 were male). Most patients had fever

(17), urticaria-like rash (18), conjunctivitis (12), oral ulcers (7), articular syndrome (14), sensorineural hearing loss (5), and 2 patients died of renal insufficiency (probably due to amyloidosis of the kidneys). The heterozygous mutations in NLRP3 have been identified in pts: T348M (3 families), R262W (2 families), A439V (1), V198M (1), Pro294Ser (1). Ten patients received canakinumab for a period of 6 months to 6.5 years, and 5 patients received anakinra before canakinumab.

Conclusion: MWS is an orphan autoinflammatory disease, however it sometimes can be observed in rheumatology practice. It is very important to acquire family medical history to identify affected family members and prescribe therapy in a timely manner. IL-1 inhibitors are an effective and safe treatment option for MWS patients.

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SAT0540 ONE-YEAR OUTCOMES AFTER RHEUMATIC IMMUNE-RELATED ADVERSE EVENTS FROM CHECKPOINT INHIBITORS

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Background: Description and initial management of rheumatic immune-related adverse-events (irAEs) from cancer immunotherapies have been reported by several groups but to date, few studies have evaluated the long-term outcomes and management of rheumatic irAEs (1).

Objectives: To describe the long-term management and assess the one-year outcomes of patients who experienced rheumatic immune-related adverse events (irAEs) due to immune checkpoint inhibitors (ICI).

Methods: This was a single-centre prospective observational study including patients referred for musculoskeletal symptoms while treated with ICI. After baseline rheumatological evaluation defining the clinical entity presented, follow-up visits were organised according to the type and severity of irAE. At one year, persistence of irAE, ongoing treatment, as well as cancer outcomes were assessed.

Results: 63 patients were included between September 2015 and June 2018. 24 patients (38%) presented with non-inflammatory musculoskeletal conditions managed with short-term symptomatic treatment and did not require specific follow-up. 39 patients (62%) experienced inflammatory manifestations, mimicking either rheumatoid arthritis (RA, n=19), polymyalgia rheumatica (PMR, n=16), psoriatic arthritis (PsA, n=3) and one flare of a preexisting axial spondyloarthritis. Overall, 32 patients (82%) received systemic glucocorticoids, with a median rheumatic dosage of 15mg/day (range: 5-60mg/day). None of the patients had to permanently discontinue ICI therapy for rheumatic irAE. 20 patients (67%) were still receiving glucocorticoids at one year, with a median dosage of 5mg/day (range: 2-20mg/day). Glucocorticoids were more frequently discontinued for patients with RA-like condition (44%) than PMR-like condition (23%), but no other predictive factor of glucocorticoids withdrawal could be identified. At one year, overall survival and progression-free survival were comparable between patients who were still receiving glucocorticoids for rheumatic irAE and patients who have discontinued. Eight patients required csDMARDs.

Conclusion: At one year, a majority of patients required long-term low-dose glucocorticoids for chronic rheumatic irAE, which seems not altering oncological control.

References:

- [1] Braaten TJ, Brahmer JR, Forde PM, et al. Immune checkpoint inhibitor-induced inflammatory arthritis persists after immunotherapy cessation. *Ann Rheum Dis*. 2019 Sep 20.

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SAT0541 THE SEVERITY OF FMF MAY BE ASSOCIATED WITH CO-MORBIDITIES

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Background: Familial Mediterranean fever (FMF) is an auto inflammatory disease with recurrent attacks of serositis. Frequent attacks and disease related sequels may be associated with co-morbidities in FMF patients.

Objectives: One of the tools for evaluating the FMF severity is the international severity scoring system for FMF (ISSF) ¹. This score includes disease related sequels, acute phase measurements, attack features and exertional leg pain.

Therefore, more severe disease may be link with subclinical inflammation, amyloidosis and frequent, prolonged and widespread attacks. All these components may augment the frequency of non-disease related co-morbidities.

Methods: We enrolled 158 FMF patients who fulfilled modified *Tel-Hashomer* Diagnosis Criteria2. The patients dichotomized based upon disease severity (mild disease or severe disease). Patients with ISSF scores lower or equal to 2 were accepted to have mild disease. Then, we compared frequency of non-disease related co-morbidities between the groups. These co-morbidities are hypertension, hypothyroidism, hyperthyroidism cardiovascular diseases, coronary artery diseases, cerebrovascular diseases, chronic renal disease (non-FMF related), chronic obstructive pulmonary diseases, and diabetes mellitus. This study was approved by the Local Research Ethics Committee and carried out in compliance with the Helsinki Declaration. All the patients gave written informed consent. P-value lower than 0.05 was considered as statistically significant.

Results: Demographic features, disease duration, smoking history and body mass index (BMI) were similar between the groups. Frequency of co-morbidity in severe disease group was statistically higher than mild disease group ($p=0.02$). Most frequent co-morbidity was hypertension in both groups.

Table. Features of mild and severe FMF groups

	Mild (n=135)	Severe (n=23)	p
Gender (M/F)	47/88	11/12	0.23
Age	36.4±11.3	36.5±14.3	0.68
Smoking (%)	38 (28.1)	5 (21.7)	0.52
BMI (kg/m ²)	24.3±9.2	24.0±8.9	0.34
Disease duration (year)	7.7±11.3	8.6±14.3	0.09
Amyloidosis (%)	2 (1.4)	3 (13.0)	0.02
Exon 10 homozygote (%)	35 (25.9)	9 (39.1)	0.19
Colchicine dosage (mg/day)	1.2±0.4	1.4±0.5	0.02
ISSF scores	0.7 ±0.7	3.4±0.5	<0.001
Co-morbidity (%)	25 (18.5)	9 (39.1)	0.02

Conclusion: In our FMF patient cohort, we found that severity of the disease may be associated with higher frequency of co-morbidities. Therefore, clinicians should be aware of the high possibility of co-morbidities in patients with more severe FMF and addressed these co-morbidities timely and properly.

References:

- [1] Demirkaya E, et al. Development and initial validation of international severity scoring system for familial Mediterranean fever (ISSF). *Ann Rheum Dis* 2016;**75**:1051-6.
- [2] Berkun Y, et al. Diagnostic criteria of familial Mediterranean fever. *Autoimmun Rev* 2014;**13**:388-90.

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SAT0542 UTILITY OF QUANTITATIVE ANALYSIS OF 18FDG-PET/CT IN IGG4-RELATED DISEASE

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Background: In IgG4-related diseases (IgG4-RD), usefulness of ¹⁸F-fluorodeoxyglucose (FDG) positron emission tomography/computed tomography (PET/CT) imaging for detecting the organ involvement of IgG4-RD have been shown and, ¹⁸FDG PET/CT was more accurate and appeared to be more sensitive as compared to other imaging technics^{1,2}. However, until now, the studies regarding about quantitative analysis of PET/CT imaging in IgG4-RD were few. To avoid unnecessary biopsy and select suitable lesion for biopsy on multi-organ involvement disease such as IgG4-RD, the information which lesion is suspected as disease-involvement lesion in a non-invasive test is important.

Objectives: The purpose of this work is to evaluate the usefulness of ¹⁸FDG-PET/CT imaging in management of IgG4-RD using quantitative analysis of PET/CT imaging.

Methods: 21 patients with IgG4-RD, in whom PET/CT was undertaken at the time of diagnosis between December 2009 and July 2018, were enrolled. We retrospectively investigated the association between histological findings in which biopsy was performed for diagnosis of IgG4-RD and findings of PET/CT. ¹⁸FDG uptake was assessed in site of major organ involvement of IgG4-RD which could be differentiated from the normal uptake of background tissue with ¹⁸FDG-PET/

CT. For quantitative analysis, we measured the highest standardized uptake value (SUV) of the pixels within the region of interest (ROI) (SUV_{max}) and the average SUV within ROI (SUV_{mean}). We also measured SUV_{mean} of liver as reference tissue. Then, we calculated ratio between SUV_{mean} of ROI and SUV_{mean}/liver.

Results: The age at diagnosis was 64.5 ± 11.9 years, serum IgG4 was 743.8 ± 584.1 mg/dl, and biopsy was performed at 24 sites (Submandibular gland 10, prostate gland 4, pancreas 2, thyroid gland 1, lung 1, retroperitoneum 1, kidney 1). Histological findings were consistent with IgG4RD (positive) at 19 sites. Although SUV_{max} at the biopsy site was not correlated with the biopsy results, SUV_{mean} at the biopsy site were significantly higher in the biopsy-positive group (figure 1). As similar, SUV_{mean}/liver SUV_{mean} were also higher in the biopsy-positive group (2.17 vs 1.52, respectively $P<0.05$). To establish cut-off value of SUV_{mean} to consider biopsy, A receiver operating characteristics (ROC) curve was constructed. ROC curve analysis indicated SUV_{mean}=4.074 as cut-off value which discriminate IgG4-RD related lesion.

Conclusion: Our present study suggested that quantitative analysis of ¹⁸FDG-PET/CT imaging is useful for selecting the biopsy site in IgG4-related disease.

Figure 1. XXXXX

References:

- [1] Ebbo M, Grados A, Guedj E, et al. Usefulness of 2-[¹⁸F]-fluoro-2-deoxy-D-glucose-positron emission tomography/computed tomography for staging and evaluation of treatment response in IgG4-related disease: a retrospective multicenter study. *Arthritis Care Res (Hoboken)* 2014; **66**(1): 86-96.
- [2] Zhang J, Chen H, Ma Y, et al. Characterizing IgG4-related disease with (1) (8)F-FDG PET/CT: a prospective cohort study. *Eur J Nucl Med Mol Imaging* 2014; **41**(8): 1624-34.

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SAT0543 PREVALENCE OF FABRY'S DISEASE IN MILD AND SEVERE FMF PATIENTS

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Background: Fabry disease (FD) is a rare metabolic disorder caused by the mutations in the α-galactosidase A (GLA) gene. FD patients present with heterogeneous clinical manifestations, which may overlap with systemic diseases including familial Mediterranean fever (FMF). Recurrent episodes of fever,