and a reduction in antibody titters against TSH receptor and anti-thyroid peroxidase. All patients were continuing treatment except for one patient who had discontinued treatment due to clinical remission. No adverse events were recorded except for mild neutropenia in one patient, which did not require dose reduction. No patient required subsequent immunosuppressive treatment or surgery.

Conclusion: Sarilumab treatment in patients with moderate-severe active OG appears to be an effective and safe therapeutic alternative, clinically similar to that obtained with TCZ in our series. This is the first published series of Sarilumab use in OG patients.

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AB1316 PREVALENCE, DIAGNOSTIC DELAY AND TREATMENT PROFILE OF PATIENTS WITH MONOGENIC AUTOINFLAMMATORY DISEASES IN AN ADULT RHEUMATOLOGY SERVICE OF A TERTIARY HOSPITAL.

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Background: Monogenic autoinflammatory diseases (MAISs) have a low prevalence and a delay in their diagnosis, with the risk of complications being AA amyloidosis the most serious. Biological therapy has been a fundamental pillar in improving the prognosis of these diseases, reducing said risk.

Objectives: We aim to study the prevalence, diagnostic delay and treatment profile of MAISs patients in an adult rheumatology service of a tertiary hospital.

Methods: Observational and retrospective study of patients with MAISs, excluding those without genetic test or negative genetic test, with a follow-up period of 17 years (2005-2021). Demographic, clinical and therapeutic variables were collected.

Results: 24 patients (50% men) were included with a mean age of 27,88 ± 11.57y. The prevalence of MAISs was 0.34%, from a total of 7,032 patients followed up in the Rheumatology service during 2020. Familial Mediterranean Fever (FMF) was the most frequent (70.83%). FMF, Tumor Necrosis Factor Receptor Associated Periodic Syndrome (TRAPS), and Hyperimmunoglobulin D Syndrome (HIDS) had a median age at symptom onset of 6, 8.5 and 2 years, a median diagnostic delay of 6, 8.75 and 11 years and a mean duration of the bouts of 4.47 (2.87), 12.25 (7.59) and 6 (2.65) days, respectively. The most frequent symptoms were: fever (79.17%), arthralgia/arthritis (79.17%) and abdominal pain (75%). During follow up 20.83% received glucocorticoids (GC) chronically (more than 3 months), 79.16% colchicine, 16.66% methotrexate and 41.66% biological therapies (Table 1). At the end of follow up 4.17% were receiving GC, 79.16% colchicine, 4.16% methotrexate and 33.33% biological therapies (Table 1). No AA amyloidosis or deaths were reported.

Table 1.

MAISs	Gender (men %)	Biological therapies prescribed during follow up (n)	Patients under biological therapies at the end of follow up (n)
FMF (n=17)	41.17%	Anakinra: 2 Canakinumab: 2 Etanercept: 1 Tocilizumab: 1	Anakinra: 3 Canakinumab: 2
TRAPS (n=4)	100%	Anakinra: 1 Canakinumab: 1 Etanercept: Tocilizumab:	Canakinumab: 1
HIDS (=3)	66.66%	Anakinra: 3 Canakinumab: 3 Etanercept. 1	Canakinumab: 2

Conclusion: MAISs prevalence was 0.34%. The diagnostic delay was of years, more than a decade for HIDS. Colchicine was widely used and well tolerated. Synthetic DMARDs had little role in treatment. Biological therapies were prescribed in a third of patients, anti IL-1 being the most used.

Disclosure of Interests: None declared

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AB1317 SERUM INTERLEUKIN 37 LEVELS IN FAMILIAL MEDITERRANEAN FEVER PATIENTS AND ASSOCIATION WITH CLINICAL FEATURES

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Background: Familial Mediterranean Fever (FMF) is the most common form of autoinflammatory diseases that is characterized by febrile episodes of serositis, arthritis and skin rash (1). Mutations in MEFV gene causes dysfunction of pyrin inflammasome, ultimately resulting in over-activation of Caspase-1 which is responsible for catalytic activation of interleukin (IL)-1 beta and gasdermin-D (2). IL-37 is also one of the IL-1 cytokines activated by caspase-1 acting as natural inhibitor of inflammation (3).

Objectives: IL-37 has pathogenetic roles for certain inflammatory diseases. We aimed to investigate serum IL-37 levels and its relationship with clinical and laboratory features of disease.

Methods: 58 adult patients diagnosed with FMF according to Tel Hashomer criteria were included. Thirty subjects were served as healthy control subjects. Demographic, genetic, clinical and laboratory features and treatment responses of patients were recorded. Twenty-nine patients were colchicine responsive whereas 29 were colchicine refractory. Serum IL-37 levels were measured by ELISA from blood samples obtained at attack free periods.

Results: Peritonitis was the most common attack type (81%) followed by fever (80%) and arthritis (67%). There was no difference between FMF patients and healthy subjects for their serum IL-37 levels. A negative correlation was found between IL-37 values and erythrocyte sedimentation rate in FMF patients (r: -0,31; p:0.015). IL -37 level was found to be significantly lower in patients who suffer from arthritis (median [IQR] 119 [396] ng/L vs 53 [164] ng/L, p= 0.03), myalgia (147 [364] ng/L vs 53 [84] ng/L, p= 0.05) or skin rash (102 [360] ng/L vs 54 [130] ng/L, p= 0.05) compared to those who did not have these attacks.

Conclusion: Although there was no difference in serum IL-37 levels between FMF patients and healthy subjects, IL-37 seem to be associated with musculoskeletal and skin attacks of FMF. Further research is needed to determine whether IL-37 have relationships with other features of FMF such as spondyloarthritis and febril myalgia.

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AB1318 ULTRASOUND EVALUATION OF ACHILLES ENTHESIS AND GASTROCNEMIUS ARCHITECTURE IN PATIENTS WITH FAMILIAL MEDITERRANEAN FEVER: PRELIMINARY RESULTS FROM A CROSS-SECTIONAL STUDY

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Background: Exertional leg pain and Achilles enthesitis are two musculoskeletal manifestations which have been described in familial Mediterranean fever (FMF) patients. Muscle performance is related both with mass and architecture. Measurements of muscle thickness and pennation angle in muscles with pennate structure, such as gastrocnemius, are representative of muscle mass and architecture, both of which can easily be measured via musculoskeletal ultrasound.

Objectives: Aim of this study is to evaluate thickness and pennation angle of medial gastrocnemius via ultrasound to evaluate muscle structure in FMF patients in order to detect implications of altered muscle condition and search for any relations with exertional leg pain/Achilles enthesitis. To our best knowledge this is the first study on the subject.

Methods: Consecutive FMF patients meeting Tel-Hashomer criteria between the ages of 18-65 were enrolled. A control group was formed from healthy volunteers

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with similar demographics. All ultrasound evaluations were performed by the same observer. Thickness and pennation angle of medial gastrocnemius was measured. Achilles tendon findings were scored according to OMERACT suggestions and total inflammation and structural damage scores recorded.

Results: A total of 40 FMF patients and 17 controls were enrolled. Clinical characteristics and ultrasound findings of subjects were presented in Table 1. All FMF patients were under colchicine treatment and 10% was also under anti-interleukin 1 treatment. Demographics were similar between groups. Right gastrocnemius pennation angle was significantly reduced in FMF group (degree, median(IQR): 20.8 (3.6) vs 22.7 (4.5), p=0.048). OMERACT ultrasound inflammation total scores were incresed on both sides in FMF group. When gastrocnemius and enthesis ultrasound findings were compared between FMF patients with and without exertional myalgia, no significant differences were observed. When FMF patients with and without enthesis findings in ultrasound examination were compared, again no significant differences in pennation angles and muscle thicknesses were observed.

Table 1. Clinical characteristics and ultrasound findings in subjects

	FMF group	Control group	
	N =40	N=17	p
Age, years, median (IQR)	42.8 (23.0)	38.0 (15.0)	0.447
Gender, female, number (%)	29 (72.5)	10 (58.8)	0.310
BMI, median (IQR)	24.6 (5.3)	24.8 (5.7)	0.485
Presence of exertional myalgia, number (%)	22 (55)		
Gastrocnemius thickness, mm, median (IQR)			
Right	19.7 (2.9)	20.7 (5.6)	0.303
Left	19.6 (2.7)	20.7 (4.6)	0.321
Gastrocnemius pennation angle, degrees, median (IQR)			
Right	20.8 (3.6)	22.7 (4.5)	**0.048**
Left	21.0 (3.3)	21.8 (4.7)	0.417
OMERACT ultrasound tissue damage total score, median (IQR)			
Right	2.0 (2.0)	2.0 (2.0)	0.659
Left	2.0 (2.0)	2.0 (2.0)	0.280
OMERACT ultrasound inflammation total score, median (IQR)			
Right	0 (2.0)	0 (0)	**0.005**
Left	0 (2.0)	0 (0)	**0.023**

Conclusion: Our preliminary results imply altered right gastrocnemius structure in FMF patients regardless of Achilles tendon involvement and exertional myalgia, which may imply altered muscle function. By increasing the number of subjects we plan to achieve more accurate results.

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AB1319 MUSCULOSKELETAL IMMUNE-RELATED ADVERSE EVENTS IN PATIENTS WITH SOLID CANCER TREATED WITH CHECKPOINT INHIBITORS: A CASE SERIES

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Background: Immune-related adverse events (irAE) are common in cancer patients treated with checkpoint inhibitors. irAE can affect any organs and systems including musculoskeletal. The characteristics of immune-related musculoskeletal disorders (MD) still remain incomplete.

Objectives: to determine the characteristics of MD manifested during the treatment of solid cancer with check-point inhibitors in case series.

Methods: We identified 13 patients with inflammatory MD manifested during treatment with check-point inhibitors. Mean age of patients was 59±10 (ranged 43-74) years, 7 (54%) of them were female. Patients were treated with anti-PD-1 (77%) and anti-PD-L1 (33%) drugs (nivolumab [n=6], pembrolizumab [n=3], atezolizumab [n=3], prolgolimab [n=1]). Cancer types include melanoma (n=5), renal cell carcinoma (n=3), bladder cancer (n=2), non-small cell lung cancer (n=1), breast cancer (n=1), cervical cancer (n=1). 7 (54%) patients had another irAE also including thyroiditis (n=3), neuropathy (n=2), rash (n=1), sicca syndrome (n=1), hepatitis (n=1). The median time of musculoskeletal manifestation after the treatment`s onset was 20 [9-48] weeks.

Results: Musculoskeletal manifestations were performed by synovitis in 9 (69%) cases, tenosynovitis in 11 (85%), entesitis in 4 (31%), morning stiffness(>30min) in 4 (31%). The most commonly affected areas were knees (77%) shoulders (69%), hands (69%) and feets (46%). Bilateral involvement was in 9 (69%) cases. MD was transient in 2 cases and persistent in 11 (9 patients with inflammatory arthritis and 2 patients with periarthritis). Clinical characteristic of inflammatory arthritis was represented by tenosynovitis more than synovitis in 3 cases (2 patients with symmetrical inflammatory polyarthritis and severe tenosynovitis, 1 patient with RS3PE-syndrome). In 6 cases inflammatory arthritis was represented by synovitis primarily (3 patients with symmetrical polyarthritis, 2 patients with oligoarthritis, 1 patient with monoarthritis). Severity of MD was assessed according to Common Terminology Criteria for Adverse Events V5.0. Grade 1 was in 2 cases (15.5%), grade 2 in 9 (69%), grade 3 in 2 (15.5%). Laboratory findings include ESR ≥30 mm/h in 7/12 (58%) cases (the median of ESR was 34 [14-42] mm/h); CRP >5 mg/l in 7/10 (70%) cases (the median of CRP was 7.2 [4.6-12.9] mg/l). RF, ACPA were negative in all cases. ANA test was positive without any specificities in 7/10 (70%) cases in dilution 1:160 (n=2), 1:320 (n=3), 1:640 (n=2). Treatment of these conditions include NSAIDs (n=10), oral systemic corticosteroids (n=5), intra-articular corticosteroids (n=1), and csDMARDs (hydroxychloroquine [n=5] methotrexate [n=1]) as corticosteroid-sparing agents. 2 patients with polyarthritis and generalized tenosynovitis and 1 patient with symmetrical polyarthritis required treatment with ≤20 mg daily of prednisone. 2 patients with poly- and oligoarthritis were treated with 5-10 mg daily of prednisone. 5 patients with arthritis required long-term treatment with NSAIDs, corticosteroids and csDMARDs (the median of follow-up was 12 [3-12] months). 1 case of the inflammatory polyarthritis with severe tenosynovitis required to suspend the anticancer treatment with checkpoint inhibitor.

Conclusion: Our data demonstrated that the musculoskeletal irAE may be represented by heterogeneous manifestation and required long-term treatment. In rare cases musculoskeletal irAE can lead to suspension of the anticancer treatment with checkpoint inhibitors.

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AB1320 JAK2 (V617F) MUTATION AND ASSOCIATION TO RHEUMATOLOGICAL DISEASES TREATMENT. STUDY OF 130 PATIENTS FROM A SINGLE UNIVERSITY HOSPITAL.

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Background: Janus Kinases (JAK) can promote cytokine production in immune and hematopoietic cells. The JAK-2 (V617F) mutation is the most frequently detected mutation in myeloproliferative neoplasms (MPN) which include essential thrombocythemia (ET), polycythemia vera (PV), primary myelofibrosis (PMF) and undifferenciated MPN. JAK-2 (V617F) mutation displays a pro-inflammatory phenotype that may be associated to a higher risk of immune mediated diseases (IMID).

Objectives: In a wide series of JAK2 (V617F) mutation, we assess the presence of a) IMID (rheumatic and non-rheumatic), b) treatment of rheumatic IMID.

Methods: We studied all the patients diagnosed with a positive JAK-2 (V16F) mutation in a single University Hospital from January 2004 to December 2019. JAK-2 (V16F) mutation was detected by using both peripheral blood and bone marrow samples. Associated IMID and treatment of rheumatic IMID were evaluated.

Results: We included 130 patients (73 men/57 women; mean age, 70.1±14.5 years). All of them were diagnosed of MPN; ET (n=64, 49.2%), PV 46 (35.4%), undifferentiated MPN (n=12, 9.2%) and PMF (n=8, 6.1%). In 10 of these 130 patients (7.7%) a rheumatic IMID was diagnosed: rheumatoid arthritis (RA) (n=4; 40%), polymyalgia rheumatica (PMR) (n=3; 30%), Sjögren syndrome (SS) (n=1; 10%), antiphospholipid syndrome (APS) (n=1; 10%) and autoinflammatory syndrome (WDR1 mutation) (n=1; 10%). Patients with RA, SS, PmR, and APS were clinically mild. RA patients were seronegative, non-erosive and without extraarticular involvement. Treatment and response are summarized in Table 1. All patients diagnosed with PMR were treated with glucocorticoids (prednisone 5 mg/12 hours), SS with hydroxychloroquine (HCQ) (200 mg/day), APS was anticoagulated with acenocumarol and one autoinflammatory syndrome was finally treated with baritinib (4 mg/day) after failure to anakinra (100 mg/day).

Conclusion: Except in autoinflammatory syndrome, most rheumatic IMID associated to JAK-2 (V16F) mutation are clinically mild, and treatment and response of patients seems similar or even better than those without this mutation.

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