were 11%, 7%, 7%, 2%, 1%, respectively. The frequency of compound mutation was 38%, and the most common compound mutation was M694V+R202Q (11%). There was significantly relationship between M694V mutation and arthritis, erysip-
pel-like erythema, proteinuria, sacroiliitis(table 2). Amyloidosis was more frequent
in patients who had M694 homozygous mutation. Mean age of disease onset was
lower in patient who had M694V homozygous mutation than M694V heterozygous
mutation ($p<0.001$).

Abstract SAT0591 – Table 1. Demografical and Clinical Features of FMF Patients

n=402	n(%)
Gender Female	241 (60)
Male	161 (40)
Fever	299 (75.5)
Abdominal Pain	347 (86)
Erysipel-like erythema	54 ⁽¹³⁾
Chest pain	83 (21)
Positive family history	174 (43)
Appendectomy	96 ⁽²⁴⁾
Amyloidosis	14 (3.5)
Chronic kidney disease	8 (2)
Inflammatory back pain	63 ⁽¹⁶⁾
Arthritis	129 (32)
Hip pain	27 (7)
Heel pain	30 (7.5)
Uveitis	4 (1)
Sacroiliitis	44 ⁽¹¹⁾
Biological treatment	16 ⁽⁴⁾

Abstract SAT0591 – Table 2. Relationship between M694V mutation and clinical findings

N(%)	Negative	Heterozygous	Homozygous	P
Fever	75 (60)	80(62)	67(75)	0,0503
Abdominal pain	107(85,6)	107(82,9)	79(88,8)	0,487
Erysipel-like erythema	5 (4)	13(10,1)	19(21,3)	0,0003
Chest pain	21(16,8)	24(18,6)	20(22,5)	0,0575
Myalgia	2(1,6)	5(3,9)	3(3,4)	0,535
Arthritis	32(25,6)	34(26,4)	41(46,1)	0,002
Sacroiliitis	11(8,8)	12(9,3)	19(21,3)	0,0096
Amyloidosis	4(3,2)	0(0)	10(11,2)	N/A
Proteinuria	7(5,6)	5(3,9)	18(20,2)	0,000

Conclusions: Tight control and sustained management are important in FMF to
protection from amyloidosis. Similar to literature, the most frequent mutation was
M694V mutation, and there was significantly relationship between M694V muta-
tion and arthritis, erysip-
pel-like erythema⁽¹⁾, proteinuria, sacroiliitis⁽²⁾ in our study.

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SAT0592 HRCT PULMONARY MANIFESTATIONS IN PATIENTS WITH SYNOVITIS, ACNE, PUSTULOSIS, HYPEROSTOSIS, AND OSTEITIS (SAPHO)SYNDROME

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Background: Synovitis, acne, palmoplantar pustulosis, hyperostosis and osteitis syndrome (SAPHO) is a rare syndrome that affects the skin, bones and joints.¹ SAPHO syndrome shares a variety of features with the seronegative spondyloarthropathies (SpAs), such as psoriatic arthritis (PsA).¹ While the extra-articular manifestations of PsA are well defined, the systemic involvements of SAPHO syndrome are only reported occasionally.² Previous cases of accompanied pleural effusion^{3, 4} and organising pneumonia⁵ were reported. In clinical work, we observed that pulmonary lesions progressed when diseases aggravated.

However, the relationship between pulmonary manifestations and diseases activity remains unclear.

Objectives: This study is to explore the pulmonary comorbidity with SAPHO syndrome.

Methods: Pulmonary HRCT images were reconstructed from whole-spine CT images in 67 SAPHO patients. HRCT images of 58 healthy controls were obtained and reviewed. Pneumonia and tuberculosis were excluded. We investigated the presence of different HRCT images findings with detailed examinations.

Results: In detailed HRCT evaluations, abnormalities were identified in 45 of all patients. We found stripe in 29 (43.3%) cases, patchy shadows in 22 (32.8%), ground-glass opacity in 11 (16.4%), pleural thickening in 9 (13.4%), solitary nodule in 6 (9%), bronchiectasis in 3 (4.5%), pulmonary bulla in 2 (3%), multiple nodules in 1 (1.5%), and interstitial change in 1 (1.5%). Compared with healthy controls, SAPHO patients have significantly higher rate of patchy shadows while significantly lower percentage of nodules (especially multiple nodules), although the overall rates of abnormal HRCT findings are similar.

Abstract SAT0592 – Figure 1. HRCT images in 3 SAPHO patients. A: Ground glass opacities (arrows); B: Solitary nodule (arrow); C: Interstitial change (arrow).

Conclusions: Our study was the first to study HRCT pulmonary changes in SAPHO patients. SAPHO patients have significantly higher percentage of patchy shadows and significantly lower rate of pulmonary nodules than healthy controls. BASDAI and age are possible good predictors for abnormal HRCT pulmonary findings.

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SAT0593 PROGRESSIVE FIBROSING INTERSTITIAL LUNG DISEASE (PF-ILD) IN PATIENTS WITH AUTOIMMUNE RHEUMATIC DISEASES

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Background: Interstitial lung disease (ILD) associated with autoimmune conditions is among the most challenging aspect of care for patients with rheumatic diseases. While idiopathic pulmonary fibrosis (IPF) is the classic fibrosing ILD, some patients with differing clinical ILD diagnoses including autoimmune associated-ILD can develop a progressive fibrosing phenotype. This phenotype is characterised by progressive pulmonary fibrosis, worsening respiratory symptoms, declining lung function, resistance to immunomodulatory therapies and early mortality. There are limited data available on current practice in diagnosis, management and treatment of PF-ILD in patients with autoimmune rheumatic diseases.

Objectives: To investigate the patient journey in patients with autoimmune rheumatic diseases and PF-ILD.

Methods: Twenty-two ILD experts from Germany, Japan, UK and the US participated in a 1 hour interview. Physicians who spend $\geq 75\%$ of their professional time managing patients and in whose caseload ≥ 10 patients had PF-ILD in the

past year completed an online survey. Physicians (243 pulmonologists, 203 rheumatologists, 40 internal medicine physicians) from the US, Japan, Germany, France, Italy, Spain and UK participated. ILD prevalence and treatment patterns in the US were analysed based on insurance claims from patients with ≥ 2 claims with an ILD diagnosis between 2014 and 2016 (ICD-9/10 codes) and ≥ 1 visit to a pulmonologist in the 3 years.

Results: Analysis of US claims identified 21 592 patients with autoimmune ILD. Rheumatoid arthritis ILD (RA-ILD) and systemic sclerosis ILD (SSc-ILD) were the most prevalent ILDs across all autoimmune rheumatic diseases. The course of the patient journey is summarised (figure 1). Most patients with autoimmune-associated ILD initially present to a rheumatologist or a primary care doctor. Both pulmonologists and rheumatologists play a key role in detection and diagnosis of ILD; however US claims data suggest that the former are more likely to make the diagnosis. Although there is significant inter-patient variation, diagnosis of ILD is estimated to take approximately 9–12 months after symptoms develop. In general ILD is diagnosed earlier in SSc. Management of patients with autoimmune-associated ILD is typically multidisciplinary, involving both a rheumatologist and a pulmonologist. The physician survey suggested that 24%–31% of patients with autoimmune-associated ILD develop PF-ILD; detection can take up to 1 year. The majority of physicians use corticosteroids as a first line treatment across autoimmune rheumatic diseases; cyclophosphamide and mycophenolate mofetil were the second and third first line option for SSc-ILD. Physicians estimate the total disease course in patients with autoimmune-associated PF-ILD to be approximately 5–7 years.

Abstract SAT0593 – Figure 1. Physician* Estimated PF-ILD Disease Course in patients with autoimmune rheumatic diseases

Conclusions: Physicians who manage patients with autoimmune diseases estimate that 24%–31% develop PF-ILD. Delayed referral to a pulmonologist or rheumatologist is likely to delay diagnosis and management of PF-ILD. Life expectancy for these patients is believed to be similar to patients with IPF. There is an unmet need for treatments that slow or stabilise disease progression of PF-ILD.

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SAT0594 WHICH ONE IS MORE VALUABLE FOR DIAGNOSIS OF ADULT ONSET STILL'S DISEASE? SOLELY NEUTROPHILIA OR LEUKOCYTOSIS WITH NEUTROPHILIA?

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Background: Adult onset Still's disease (AOSD) is a rare, auto-inflammatory disease that has unknown etiology and poorly defined pathogenesis. To date, there are several classification criteria, available for AOSD, but most commonly used one is proposed by Yamaguchi et al.¹ One of the major items of this set of criteria is leukocytosis with neutrophilia.

Objectives: To usefulness of solely neutrophilia instead of leukocytosis with neutrophilia for the Yamaguchi criteria.

Methods: Sixty-one AOSD patients followed at Hacettepe University department of rheumatology were included to analysis. Diagnosis of AOSD was based on physicians' decision, but Yamaguchi criteria was applied for all patients, as well. Complete blood test with differential was noted at the time of diagnosis of AOSD. One of the major criteria of Yamaguchi criteria was "leukocytosis with neutrophilia (defined as more than 80% of leukocytes would be granulocyte)". Leukocytosis [$>10000/\text{mm}^3$ x1000] and neutrophilia [(6400/ mm^3) x1000] were defined according to normal upper limits of our laboratory. To compare with Yamaguchi criteria, neutrophil/leukocyte ratio for all patients were calculated. Descriptive statistics for non-normally distributed countable data were given as median and interquartile range (Med [IQR]). $p < 0.05$ was considered as statistically significant.

Results: Total 61 patients (46 (75%) female) were recruited. Prevalence of major and minor Yamaguchi criteria were as follow: fever in all patients, arthralgia in 58 (95%) patients, rash in 43 (71%) patients, sore throat in 48 (79%) patients, lymphadenopathy and/or splenomegaly in 23 (38%) patients, absence of RF and ANA in 58 (96%) patients, elevated liver enzymes in 47 (77%) patients. Median leukocyte and neutrophil count were 13.5 (IQR;10.1–18.9) x 1000/ mm^3 , 12 (IQR;7.2–17.6) x 1000/ mm^3 , respectively. "Neutrophilia $> \text{UNL}$ " was significantly more prevalent than "leukocytosis with neutrophilia $> 80\%$ " (51 (83.6%) vs. 37 (60.6%), $p < 0.001$). Overall, 14 (23%) patients would have one more positive major criteria if "neutrophilia $> \text{UNL}$ " would be used instead of "leukocytosis with neutrophilia $> 80\%$ ". When current Yamaguchi criteria (including leukocytosis with neutrophilia $> 80\%$) applied to patients with AOSD, 57 (93.5%) of 61 patients were met the criteria. However, when "neutrophilia $> \text{UNL}$ " was used instead of "leukocytosis with neutrophilia $> 80\%$ ", all patients were met the revised criteria.

Conclusions: As all patients who had leukocytosis also had neutrophilia but it was not vice versa. In other words, choosing "leukocytosis with neutrophilia $> 80\%$ " instead of "neutrophilia $> \text{UNL}$ " can underdiagnose AOSD approximately in 7% of patients according to Yamaguchi criteria. Moreover, Physicians should be keep in mind that reactive hemophagocytosis may influence the count of leukocyte. Consequently, using "neutrophilia $> \text{UNL}$ " as a criteria instead of "leukocytosis with neutrophilia $> 80\%$ " may be more appropriate for the diagnosis of AOSD in real life.

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SAT0595 ANALYSIS OF RECURRENCES AFTER SUSPENSION OF IMMUNOSUPPRESSIVE TREATMENT IN NON-INFECTIOUS UVEITIS

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Background: Uveitis is the most common ocular inflammatory disease. According to the classification, non-infectious uveitis 70% of the total, presenting a chronic course and with it, an increased risk of complications. The phased therapeutic approach in ophthalmology is well established and is based on the efficacy of synthetic and biological immunosuppressive therapy.¹ However, we do not have evidence-based protocols that allow us to know how long to maintain them, or when to interrupt them.

Objectives: The main objective is to evaluate the free time without ocular inflammatory activity after the withdrawal of the synthetic and/or biological immunosuppressive treatment in patients with non-infectious uveitis.

As secondary objectives are collected the epidemiological and clinical characteristics, the distribution of the different immunosuppressive drugs employed and