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, erysipel-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)
İnflammatory 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
Miyalgia	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 mutation and arthritis, erysipel-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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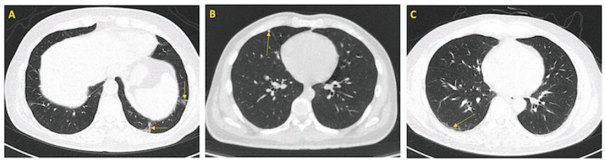
Background: Synovitis, acne, palmoplantarpustulosis, hyperostosis and osteitis syndrome (SAPHO) is a rare syndrome that affects the skin, bones and joints.[1] SAPHO syndrome shares a variety of features with the seronegative spondyloarthropathies (SpAs), such as psoriatic arthritis (PsA).[1] While the 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 pneumonia([5were 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 patientsHRCT 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 ≥75% of their professional time managing patients and in whose caseload ≥10 patients had PF-ILD in the