the group was 28±8 (range 20-40). All patients diagnosed with IRP were negative for autoinflammatory genetic screening, while a MEFV variant (K695R het.) was detected in the FMF patient. Median duration of follow-up was 30 months (range 11-129). In table 1, demographic and clinical features are given. The median number of recurrence was 6 before anakinra treatment. No episode of pericarditis was observed in any of the patients after the initiation of anakinra. The response to anakinra persisted even after the dose was reduced to 100 mg/alternate day in 3 patients, however in 2, recurrence of pericarditis was observed and anakinra was escalated to initial dose. It was possible to discontinue corticosteroid treatment in all patients. Currently all patients continue anakinra treatment. No side effect including injection site reaction, has been observed by now.

Conclusion: Anakinra seems to be a safe and effective treatment approach for colchicine resistant recurrent pericarditis. However recurrence may occur during dose tapering.

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FRI0617 APPLICATION OF AUTO-INFLAMMATORY DISEASE DAMAGE INDEX (ADDI) TO PATIENTS WITH FMF AND RELATED FACTORS WITH DAMAGE

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Background: Familial Mediterranean Fever (FMF) is the most frequent auto-inflammatory disease caused by MEFV gene mutations. Available reports investigated only specific components of damage such as amyloidosis. All possible organ targets of damage have not been entirely evaluated before. Such as Disease severity index which is emerged especially for FMF do not cover entire damage domains related to FMF. Recently, a new scoring system (Auto-inflammatory disease damage index) was developed and validated for autoinflammatory diseases.

Objectives: We aimed to investigate damage accrual caused by FMF and associated features with damage.

Methods: All patients recruited from FMF in Central Anatolia (FICA) cohort, which is a duplication disabled, internal and externally controlled, cross-sectional, multicenter accessible web-based cohort. This study is comprising 970 adult patients (mean age 35.3 ±12.1 years, 61.5% female). Demographic data, FMF disease characteristics, co-morbid conditions, disease complications were meticulously questioned and laboratory features and genotype data (if available) were recruited from patient files. FMF caused damage was assessed by auto-inflammatory disease damage index (ADDI) which is recently validated. Association between damage and demographic, disease and treatment characteristics were analyzed.

Results: Proportions of FMF manifestations were fever 83.1%, peritonitis 91.5%, pleuritis 47.9%, arthritis 43.3% and skin rash 26.2%. Dominant attack types were fever in 6.2%, serositis in 65.7%, musculoskeletal in 16.8% and all types of attacks were common in rest of patients. MEFV mutations were available in 814 subjects and 75.9% of these subjects were harboring M694V mutation (42.5% homozygous for M694V). Among all 63.1% patients were well responded to colchicine and 8.8% were non-responders. Median ADDI score was 1 (min 0-max 11). Most common FMF related damages were observed in musculoskeletal, reproductive and kidney domains. Chronic musculoskeletal pain was present in 49%, joint deformity in 2.9%, infertility in 6.6%, amenorrhea in 3.9%, proteinuria in 6.9%, amyloidosis in 5.9% and renal failure in 3.7% of the patients. 411 (%42.3) of patients had no damage accrual. M694V homozygous mutation, male gender and colchicine nonresponse were found to be the independent predictors of damage.

Conclusion: M694V homozygous mutation, colchicine non-response and male gender are predictors of damage and effective therapeutic interventions must be undertaken to prevent from damage in these patients.

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FRI0618 ANALYSIS OF NEW REFERRALS TO A SPECIALIST UK ADULT AUTOINFLAMMATORY DISEASE SERVICE

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Background: Diagnosis of the systemic autoinflammatory diseases (SAIDs) requires a high index of suspicion and previous series has suggested that there are often long diagnostic delays, particularly in TRAPS and MKD.

Objectives: To look at the case mixed referred to a single adult clinic in London specialising in assessment of potential SAIDs over the course of the year of 2017.

Methods: All new referrals were accepted for clinical assessment. At the first visit patients had a full history and examination, genetic testing – varying from single gene to a 20 gene panel depending on clinical features, and laboratory testing including fortnightly blood draws for serial analysis of the hepatic acute phase response proteins, CRP and SAA over a 3 month period.

Patients with a non suggestive history, non contributory genetic testing and no evidence of inflammation accompanying symptoms were felt not to have SAIDs and referred back to their local hospitals for further management. Other cases were diagnosed based on full clinical assessment, other investigations – for example ferritin in AOSD, genetic testing results, serial monitoring of CRP and SAA and therapeutic trials, for example colchicine in presume FMF and anti IL-1 therapies in CAPS and Schnitzler's syndrome

Results: 273 new patients were referred. Median age at referral was 37.4 years, the oldest patient was 84.3 years old and 59% were female. 174 (64%) were of northern European ancestry, 68 (25%) were eastern Mediterranean, west Asian or southern European ancestry, 19 (7%) were of south or east Asian ethnicity and 4% were of African or Afro-Caribbean ancestry. 76% of referrals were from hospital specialities. The referral source was: rheumatology 38%, general practitioner 24%, dermatology 8%, immunology 8%, gastroenterology 6%, infectious diseases 3%, clinical genetics 3%, nephrology 2%, haematology 2%, gynaecology 2%, emergency department 1%, respiratory 1%, other 2%.

After work up 135 (49.5%) were felt not to have a SAID as the cause of their symptoms. Of the remaining 138 patients who did have evidence of a SAID the diagnoses made were: FMF 33%, uncharacterised SAID 26%, CAPS 9%, AOSD 8%, recurrent idiopathic pericarditis 6%, Schnitzler's Syndrome 5%, TRAPS 4%, variant PFAPA 4%, DADA2 1%, MKD 1%, CRMO 1%, Behcets 1%, Cattleman's disease 1%.

The median interval between reported symptom onset and diagnosis were as follows: 16 yrs for FMF, 28.1 yrs for CAPS, 5.0 years for recurrent idiopathic pericarditis, 4.5 yrs for Schnitzler's Syndrome, 5.7 yrs for TRAPS, 20.5 yrs for variant PFAPA, 12.5 yrs for DADA2, 17 yrs for MKD and 2 years for CRMO.

Conclusion: This series suggests that recognition and diagnosis of the SAIDs remains a challenge. More than 1/3 of referrals were from rheumatology, referrals from primary care were almost exclusively from patients with a known family history of one the inherited syndromes. The wide variety of referring specialities reflects the diverse nature of SAIDs and the importance of almost all specialities considering the possibility of SAIDs. Only just over 50% referrals had evidence of diseases falling within the recognised SAID spectrum and 26% of these have currently uncharacterised disease with no diagnostic genetic testing. Of those in whom a diagnosis could be made there are significant diagnostic delays fortunately despite late initiation of treatment no patients had evidence of systemic AA amyloidosis.

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FRI0619 INPATIENT BURDEN AND COMORBIDITIES OF SARCOIDOSIS: NATIONWIDE INPATIENT SAMPLE 2013–2014

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Background: Little is known about the inpatient burden and healthcare utilization among patients with sarcoidosis. Previous studies have focused on trends of hospitalization rate of patients with sarcoidosis over time [1] but none has investigated their inpatient prevalence, mortality and expenditures.

Objectives: The current study was conducted with the aims to shed more light on those characteristics as well as to investigate the comorbidities of sarcoidosis using the data from a large national database.

Methods: Patients with sarcoidosis were identified within the Nationwide Inpatient Sample (NIS) database of the years 2013 and 2014 using the ICD-9 diagnostic code. NIS is the largest publicly available inpatient database in the US. Data for more than seven million individual hospitalization across all-payers in the United States (US) is recorded annually in the NIS database. This is itself a 20% stratified sample of over 4,000 non-federal acute care hospitals from more than 40 states of the US and is representative of 95% of hospital discharges nationwide. Data on patient and hospital characteristics, comorbidities, total hospital costs and total hospitalization charges was collected. A propensity-matched cohort of patients without sarcoidosis from the same database was created and used as comparators for the analysis of comorbidities. Inpatient prevalence of sarcoidosis was calculated using all admissions in the NIS database as denominator. Odds ratios (OR) comparing the prevalence of comorbidities between cases with sarcoidosis and propensity-matched controls without sarcoidosis were calculated.

Results: A cohort 78,055 patients with sarcoidosis was identified from the database, corresponding to an inpatient prevalence of 2.21 cases per 1,000 admissions. The most common reasons for admission among patients with sarcoidosis in this cohort were as follows; pneumonia (34.3%), respiratory failure (26.1%), cardiomyopathy (10.9%), coronary artery disease (9.2%), acute kidney injury (8.5%) and atrial fibrillation (5.5%). Analysis of comorbidities found that patients with sarcoidosis had significantly higher odds of atrial fibrillation, conduction abnormalities, aortic valvulopathy, congestive heart failure and cardiomyopathy compared to propensity-matched patients without sarcoidosis (table 1). After adjusting for confounders, patients with sarcoidosis displayed a mean additional \$27,205 (p<0.01) for total hospitalization charges (the amount of money that each hospital billed for providing its service on each case) when compared to hospitalization of patients without sarcoidosis.

Conclusion: The inpatient prevalence of sarcoidosis was relatively high compared to its overall incidence. Hospitalization of patients with sarcoidosis was associated with a significantly higher total hospitalization charges. Patients with sarcoidosis have higher risk of several cardiac comorbidities.

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Table 1. Odds ratio comparing comorbidities between patients with sarcoidosis and patients without sarcoidosis

Variable	Adjusted odds ratio	95% Confidence Interval	p-value
Atrial fibrillation	1.41	1.13-1.76	<0.01
Conduction abnormalities (blocks)	2.04	1.45-2.89	<0.01
Coronary artery disease	1.10	0.94-1.29	0.26
Congestive heart failure	1.23	1.04-1.45	0.02
Cardiomyopathy	1.25	1.08-1.44	<0.01
Aortic valvulopathy	1.78	1.30-2.44	<0.01

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FRI0620 FMF PATIENTS HAVE A HIGHER PREVALENCE OF SPA-RELATED DISORDERS INCLUDING MHC-I-OPATHIES THAN CONTROLS: INSIGHTS FROM A LARGE COHORT STUDY

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Background: Familial Mediterranean fever (FMF) is a common, hereditary autoinflammatory disorder, caused by mutations in the MEFV gene encoding for pyrin. MEFV mutations have been also reported in spondyloarthritis (SpA)-associated disorders including Crohn's disease, ulcerative colitis, Behçet's disease, psoriasis and ankylosing spondylitis (AS) with the latter 3 disorders considered under the umbrella term of "MHC-I-opathies" due to population-level associations of HLA-B51, Cw0602 and HLA-B27, respectively. FMF and the SpA group of disorders share an association with disease localization to site of physical stress or microdamage with innate immune activation at such sites as a primary driver of immunopathology but this is not the case for classical autoimmune diseases where central tolerance failure is an overarching immunological concept.

Objectives: To test the hypothesis that autoinflammation in FMF may exaggerate the tendency towards adaptive immunopathology or MHC class-I associated disorders and therefore a higher prevalence of SpA-related disorders in patients with FMF.

Methods: 7,747 FMF patients and 10,080 age- and sex-matched controls in the Clalit-Health-Services medical database were identified and compared in terms of prevalence of SpA-associated disorders and also evaluated 4 classical and strong MHC class-II associated disorders, namely sarcoidosis, pernicious anaemia, pemphigus vulgaris, and myasthenia gravis to ascertain whether such associations with SpA-spectrum disease were specific or merely reflected the non-specific consequences of innate immune system activation on driving divergent types of immunity.

Results: FMF showed a strong association with MHC class I related diseases: OR 28.58 ([95%CI 6.93-117.87], p<0.0001) for Behçet's disease, OR of 10.33 ([95%CI 4.09-26.09], p<0.0001) for AS, and OR 1.67 ([95%CI 1.19-2.33], p=0.0029) for psoriasis. For weakly MHC class I linked diseases, an OR of 3.76 ([95%CI 2.48-5.69], p<0.0001) for Crohn's disease and OR of 2.64 ([95%CI 1.52-4.56], p=0.0005) for ulcerative colitis were found. No association was found between FMF and strongly MHC class II-associated including pemphigus vulgaris, sarcoidosis, pernicious anaemia and myasthenia gravis. At the Cox multivariate survival analysis, the mortality of FMF patients was higher only in Crohn's disease with an HR of 2.32 ([95%CI 1.09-4.93], p=0.0291).

Conclusion: FMF patients are associated with increased risk of SpA-related disease diagnosis including MHC-I-opathies suggesting that tissue-specific dysregulation of innate immunity share between FMF and SpA spectrum disorders may drive adaptive immune MHC class I associated conditions.

Figure 1. The proportion of FMF patients with SpA related disorders (A) and their impact on FMF patients' survival (B)