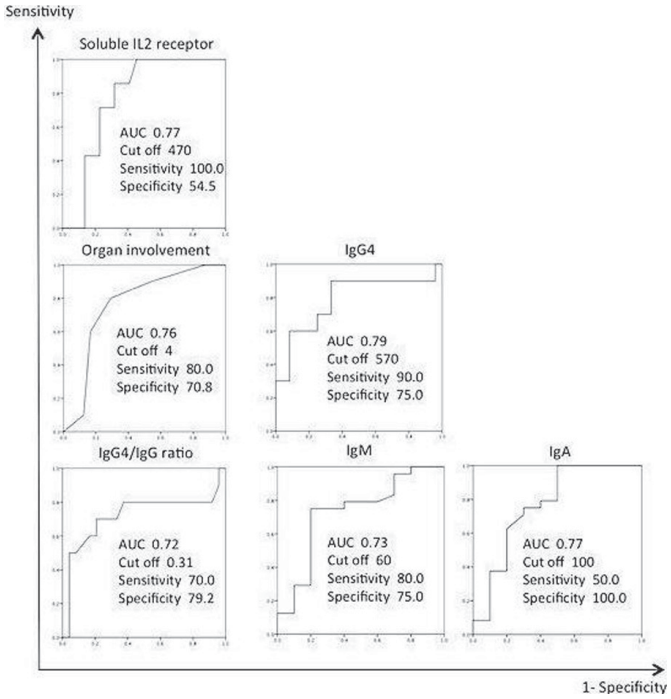

Figure 1

the higher likelihood of disease relapse following GCs therapy in patients with IgG4-RD.

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FRI0592 CLINICAL, ANALYTICAL AND RADIOLOGICAL CHARACTERISTICS IN A COHORT OF PATIENTS WITH SARCOIDOSIS

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Background: Sarcoidosis is a systemic granulomatous disease, frequently affecting lungs, eyes and skin, although it may damage other organs, among them the musculoskeletal system

Objectives: To describe the clinical characteristics and radiological pattern in a cohort with predominantly pulmonary sarcoidosis, and to determine the relationship between the levels of angiotensin converting enzyme (ACE), pulmonary radiological stage and sarcoidosis course (chronification or remission)

Methods: Data from 2328 patients in an Interstitial Lung Diseases consultation during the first half of 2016 were analyzed. Out of these, 50 had sarcoidosis.The delay in the diagnosis of sarcoidosis was defined as the difference in years between the diagnostic suspicion and diagnosis of sarcoidosis.

Chi square tests were used, assuming an error of the first species not higher than 0.05, in order to: 1. Study the association between angiotensin converting enzyme (ACE) levels and binary variables (extrapulmonary symptoms, radiological stage and evolution of S) 2. Determine the association between evolution, the radiological stage and the presence of extrapulmonary symptoms.

Results: We included 29 (58%) women and 21 (42%) men, (mean age of 44±11.7 years). Initial diagnosis: 88% S, 8% lymphoma and 4% tuberculosis. Of the 44 diagnosed cases of S, 24 were on the first visit, 11 the following year and 1 seven years later. Of the 4 lymphomas, 2 were diagnosed of S that same year and the other 2 were diagnosed the following year. Of the 2 tuberculosis, one was diagnosed of S in one year and the other at 4 years.

The most frequent extrapulmonary manifestations were cutaneous 24%, followed by the articular, cardiac and ocular in 10%, neurological 8% and renal 4%. In 6% of patients, the first clinical manifestation of the disease was bilateral arthritis of the ankles, The ACE title is increased in 62% of patients, normal in 34%. The mean and standard deviation of the title of patients with an increased ACE value was 150.5 and 53.4 IU/L, respectively. In all patients, x-ray and high resolution tomography were performed, with stage 2 being the most frequent (44%), followed by 3 (20%), 0 and 1 (14%) and 4 (8%). Histological confirmation was obtained by transbronchial biopsy (66%), cutaneous (12%) or lymph node biopsy (12%) in 90% of the patients. 90% of patients have been treated with oral glucocorticoids and 42% associate immunosuppressive therapy.

The ACE levels showed no statistical association with any of the variables studied, although a very clear association (p=0.04754) was observed between the course of the disease and the presence of extrapulmonary symptoms: from the 25 patients without extrapulmonary symptoms, only in 35% of cases the process become chronic.

Conclusions: Our results, in general, coincide with what is published in the literature. In our cohort, initial diagnosis of S was relatively high (28/50 =56%), while misdiagnosis was relatively low (6/50 =12%). The level of ACE does not seem to be clearly associated with the presence of extrapulmonary symptoms, nor with the course of S. However, the presence of extra-pulmonary symptoms seems to lead to a chronification

Disclosure of Interest: None declared

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FRI0593 ASSOCIATION OF THYMOMA WITH AUTOIMMUNE DISEASES IN A SERIES OF 83 CASES

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Background: Thymoma is the most common neoplasm originated from the thymus gland and accounting for 50% of anterior mediastinal tumors. Within its clinical manifestations are included the loss of self-tolerance and the development of autoimmunity.

Objectives: To study the frequency of autoimmune diseases (AD) in patients with thymoma and to describe their clinical characteristics and outcome.

Methods: We performed a retrospective observational study of a cohort of patients diagnosed with thymoma and followed-up in our center between January 1985 and September 2016. The variables evaluated included demographics, thymoma characteristics, clinical and analytical manifestations of autoimmunity, treatment and outcome.

Results: A total of 83 patients were included, 56.6% of them women, with a mean age at diagnosis of the thymoma of 58.4±15.8 years (range: 16–94), 31.3% of which corresponded to type I The classification of Masaoka, 39.1% to II, 17.2% to III and 10.9% to IV. There were one or more AD associated in 41 cases (49.4%). The most frequent diagnoses were myasthenia gravis (19), systemic lupus erythematosus (SLE) (4), subacute cutaneous lupus erythematosus (1), Sjögren's syndrome (1), rheumatoid arthritis (1), spondyloarthritis (1), sarcoidosis (1), hemolytic anemia (2), pernicious anemia (1), aplastic anemia (1), cutaneous limited systemic sclerosis (1), urticaria-vasculitis, erythroblastopenia (1), recurrent pericarditis (1), thyroid disease (2) and lichen planus (1). The diagnosis of AD preceded to thymoma in 38.2% of cases and was later in the remaining cases. In 4 cases there was also a concomitant primary immunodeficiency (variable common immunodeficiency 3, CD4 immunodeficiency 1). The most frequently identified autoantibodies were anti-acetylcholine receptor (14/41, 34.1%), anti-striated muscle (3/41, 7.3%), ANA (11/41, 26.8%), (3/41, 7.3%), rheumatoid factor (3/41, 7.3%), anti-thyroid (3/41, 7.3%), antiphospholipids (2/41, 9%) and anticentromere (1/41, 2.4%). In the comparison of patients with and without associated AD, no significant differences were found regarding age, sex or Masaoka classification. There were 6 deaths, 4 in group with associated AD and 2 in the group without AD, but without significant difference (p=0.3797).

Conclusions: In the analyzed population of patients with thymoma of our center, almost half of them developed AD, which in a major group preceded the diagnosis of neoplasia. The spectrum of autoimmunity associated with thymoma was quite broad, including organ-specific AD such as myasthenia gravis (which is most frequently described in the literature) and autoimmune cytopenias, but also to systemic AD, the most common being SLE. The autoimmunity study should be included in the assessment of the patient with thymoma as it could contribute to the early diagnosis of associated AD.

Disclosure of Interest: None declared

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FRI0594 A CASE OF MOSAICISM IN TNF ASSOCIATED PERIODIC SYNDROME (TRAPS)

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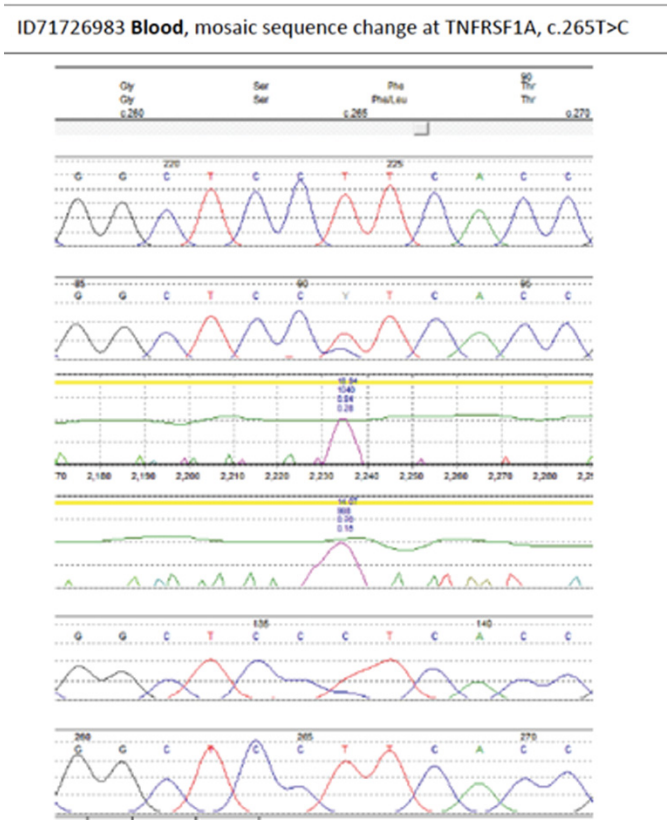
Background: Tumor necrosis factor receptor (TNFR)–associated periodic syndrome (TRAPS) is an autosomal-dominant disease caused by gain-of-function mutations in the TNFRSF1A gene, which encodes the 55-kd TNFR type I (TNFRI) protein. Mosaicism has been recently idenitfied in a single patient.[1] A 60 year old male presented with a 6 year history of intermittent fever as high as 103.5, lasting 3–4 weeks with associated peritoneal symptoms, arthralgias, myalgias, lymphadenopathy, bilateral episcleritis, erythematous rash in his torso. Prednisone up to 60 mg daily only partially alleviated his symptoms and colchicine was ineffective.

Objectives: To explore the role of mosaicism in a patient with adult onset TRAPS phenotype.

Methods: DNA was extracted from the patient's whole blood, saliva and hair root. The TNFRSF1A gene was analyzed by Sanger sequencing in all tissues, and next-generation sequencing in whole blood. In silico molecular modeling was performed to predict the structural and functional consequences of the tumor necrosis factor receptor (TNFR) type I protein mutation.
Results: Sanger sequencing and next-generation sequencing methods revealed differential tissue presence of a misense mutation at c.265 T>C p.Phe89Leu (F89L) Chr12(GRCh37):g.6442960A>G ex3 rs104895245. The mutant allele was present in whole blood and buccal mucosa and absent in hair root, supporting the presence of somatic TNFRSF1A mosaicism. In silico prediction modeling with SIFT and PolyPhen2 suggested that this mutation led to numerous structural rearrangements which resulted in changes in the protein surface profile. The patient had a complete response to treatment with canakinumab an interleukin-1 beta blocker, with resolution of symptoms and normalization of acute-phase protein levels.



Conclusions: This is the second reported case of TNFRSF1A mosaicism in a patient with TRAPS, which was attributable to a de novo mosaic missense mutation in the TNFRSF1A gene. (c.265 T>C) p.Phe89Leu (F89L) The clinical picture in this patient, including the complete response to IL-1 blockade, was typical of that found in TRAPS. This case suggests that adult onset TRAPS phenocopies may be attributed to somatic (or postzygotic) mutations.
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[1] Rowczenio et al Association of Tumor Necrosis Factor Receptor–Associated Periodic Syndrome With Gonosomal Mosaicism of a Novel 24-Nucleotide TNFRSF1A Deletion ARTHRITIS & RHEUMATOLOGY, Vol. 68, No. 8, August 2016, pp 2044–2049.
Disclosure of Interest: None declared
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FRI0595 EPIDEMIOLOGY AND COMPLICATIONS OF HOSPITALIZED PATIENTS WITH ADULT ONSET STILL'S DISEASE IN UNITED STATES: A NATIONWIDE ESTIMATE

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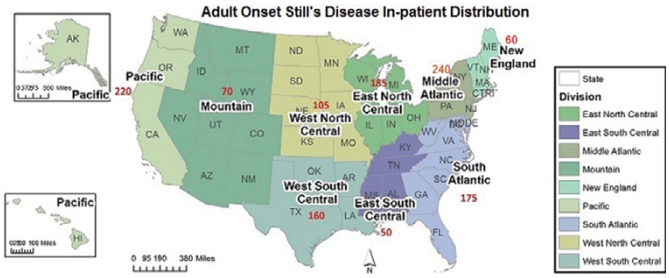
Background: There is a dearth of epidemiological studies in the US and worldwide on Adult Onset Still's Disease (AOSD). Currently, there is no consensus on its incidence and prevalence in different populations. Most studies report a majority of patients below the age of 35.[1]
Objectives: To describe the demographics, complications and mortality of hospitalized patients with AOSD in United States.
Methods: All adult (>18 years) hospitalized patients in 2013 from a nationwide inpatient sample (NIS) database were captured. AOSD patients were identified using the ICD-9 code 714.2 that was in use in 2013. Patients also coded for Rheumatoid Arthritis, Lupus, Myositis, Polymyalgia Rheumatica, Ankylosing Spondylosis and Psoriatic Arthritis were excluded. This was done in order to truly capture patients with strictly AOSD. NIS is the largest all- payer inpatient care database in the United States with approximately 8 million hospitalizations each year. Discharge weights were used to enable nationwide estimates. Descriptive statistics were represented as means/medians for continuous and as frequencies and percentages for categorical variables.
Results: In 2013, 1410 US patients were coded with the ICD-9 code 714.2 and, after excluding concomitant rheumatic disease diagnoses as per protocol, 1265 AOSD patients were analyzed. AOSD patients had a mean age of 53.8 (SD - 18.1) years and a median age of 54 years. 70.4% were females. The racial/ethnic distribution showed that 61.6% white, 13.9% African American and 17.3% Hispanic patients were affected. 56.9% were hospitalized in urban teaching hospitals. The Mid-Atlantic census division had the highest number of patients – 240 (Figure). 35 (2.8%) of patients developed Macrophage Activating Syndrome (MAS). 30 (2.4%) patients had other severe complications of AOSD like disseminated intravascular coagulation (DIC) and thrombotic thrombocytopenic purpura (TTP). Median length of stay was 4 days and median hospital charges were $31,400 (Table). There were 35 inpatient deaths (mortality 2.8%): 71.4% of deaths were in females, 50% in Asians, and 71.4% were in patients in the 54- 67 age group.

Variables	N=1265	Percentage
Age (mean age in years (standard deviation))	53.9 (18)	
Female Sex	890	70.4%
White	730	61.6%
Black	165	13.9%
Hispanic	205	17.3%
Asian	50	4.2%
Bed Size of the Hospital		
Small	140	13.7%
Medium	270	26.5%
Large	610	59.8%
Location/teaching status of hospital		
Rural	100	9.8%
Urban non-teaching	340	33.3%
Urban teaching	580	56.9%
Macrophage Activating Syndrome	35	2.8%
Other complications - (DIC and TTP)	30	2.4%
In-hospital death	35	2.8%
Length of stay (Median no of days)	4 days	
Total Hospital Charges (Median in dollars)	$ 31,400	



Conclusions: In hospitalized American AOSD patients, the average age was higher than previously thought. This may indicate an aging population with a higher number of comorbidities that justify hospitalization. Mortality increased with age and was higher among women and Asians. To our knowledge, this is the largest epidemiological study of AOSD today in the USA.
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[1] Sakata N, Shimizu S, Hirano F, Fushimi K. Epidemiological study of adult-onset Still's disease using a Japanese administrative database. Rheumatol Int 2016;36(10):1399–405 doi: 10.1007/s00296–016–3546–8.
Disclosure of Interest: None declared
DOI: 10.1136/annrheumdis-2017-eular.5490

FRI0596 ANAKINRA TREATMENT IN PATIENTS WITH FAMILIAL MEDITERRANEAN FEVER: A SINGLE-CENTER EXPERIENCE (CASE SERIES)

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Background: Approximately in 5 to 10% of FMF patients there is insufficient response and/or intolerance to colchicine treatment. Several reports have pointed out the efficacy of IL-1 blockade in colchicine resistant FMF subgroup.
Objectives: To review the patients followed in our center with FMF who received Anakinra, an anti-IL-1 receptor antagonist, in terms of outcome and side effects.
Methods: 43 FMF patients who were treated with Anakinra were retrospectively