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 missense 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.

ID71726983 Blood, mosaic sequence change at TNFRSF1A, c.265T>C

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.

References:

[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
DOI: 10.1136/annrheumdis-2017-eular.3485

FRI0595 **EPIDEMIOLOGY AND COMPLICATIONS OF HOSPITALIZED PATIENTS WITH ADULT ONSET STILL'S DISEASE IN UNITED STATES: A NATIONWIDE ESTIMATE**

B. Mehta¹, W. Briggs², P. Efthimiou^{3,4}. ¹Rheumatology, HSS; ²New York Presbyterian/ Brooklyn Methodist Hospital, New York, United States; ³Rheumatology, New York Presbyterian/ Brooklyn Methodist Hospital; ⁴Medicine, Weill Cornell Medicine, New York, United States

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 Spondylitis 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.

References:

[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)**

B. Ergezen, S. Ugurlu, H. Ozdogan. Division of Rheumatology, Department of Internal Medicine, Cerrahpasa Medical Faculty, University of Istanbul, Istanbul, Turkey

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