Giovanna Fabio: None declared, Laura Obici: None declared, Maria Alessio: None declared, Donato Rigante: None declared, Luca Cantarini: None declared, Antonella Insalaco: None declared, Marco Cattalini: None declared, Maria Cristina Maggio: None declared, Gabriele Simonini: None declared, Alma Nunzia Oliveri: None declared, Serena Pastore: None declared, Maddalena Lancieri: None declared, Nicolino Ruperto Grant/research support from: Bristol-Myers Squibb, Eli Lilly, F Hoffmann-La Roche, GlaxoSmithKline, Janssen, Novartis, Pfizer, Sobi (paid to institution), Consultant of: Ablynx, AbbVie, AstraZeneca-Medimmune, Biogen, Boehringer Ingelheim, Bristol-Myers Squibb, Eli Lilly, EMD Serono, GlaxoSmithKline, Hoffmann-La Roche, Janssen, Merck, Novartis, Pfizer, R-Pharma, Sanofi, Servier, Sinergie, Sobi, Takeda, Speakers bureau: Ablynx, AbbVie, AstraZeneca-Medimmune, Biogen, Boehringer Ingelheim, Bristol-Myers Squibb, Eli Lilly, EMD Serono, GlaxoSmithKline, Hoffmann-La Roche, Janssen, Merck, Novartis, Pfizer, R-Pharma, Sanofi, Servier, Sinergie, Sobi, Takeda, Marco Gattorno Consultant of: Sobi, Novartis, Speakers bureau: Sobi, Novartis

DOI: 10.1136/annrheumdis-2020-eular.1402

OP0274

CLINICAL ASPECTS, LABORATORY CHARACTERISTICS AND TREATMENT RESPONSES OF AA AMYLOIDOSIS: SINGLE CENTER EXPERIENCE WITH 163 PATIENTS

M. Bektas¹, N. Koca¹, E. Oguz², B. Ince¹, Y. Yalçinkaya¹, B. Artım-Esen¹, M. Inanc¹, M. L. Ocal¹, A. Gül¹. ¹Istanbul Faculty of Medicine, Internal Medicine, Rheumatology Division, Istanbul, Turkey; ²Istanbul Faculty of Medicine, Internal Medicine, Rheumatology Division, Istanbul, Turkey

Background: AA amyloidosis has been associated with uncontrolled chronic inflammatory diseases such as rheumatoid arthritis (RA), ankylosing spondylitis (AS), inflammatory bowel disease (IBD) and hereditary periodic fever syndromes, and the most common cause is familial Mediterranean fever (FMF) in Turkey.

Objectives: We herein aimed to evaluate clinical and laboratory characteristics and treatment responses of patients with AA amyloidosis retrospectively in a tertiary referral center.

Methods: Study group was consisting of patients with biopsy proven AA amyloidosis, and their data were recorded from their charts. Treatment responses were categorized as follows: *complete response* was defined as no increase in serum creatinine and a proteinuria below 1gr/day; *partial response* as 50% decrease in proteinuria; and *stable disease* as no significant change in serum creatinine and proteinuria. Progressive disease was defined as increase in serum creatinine and/or proteinuria under treatment.

Results: 173 patients were identified, and 10 patients with no biopsy result and/or missing data were excluded. A total of 163 patients (79 females, 84 males) were included in the study. Median age of patients was 45.4, and median age at diagnosis of amyloidosis was 33.5. Most common cause of amyloidosis was FMF (78.5%), followed by idiopathic cases (7.9%) and patients with AS (4.9%). A quarter (26%) of amyloidosis patients had a family history for AA amyloidosis, and 59% of patients with FMF had a family history of FMF. Amyloidosis was confirmed by renal biopsy in 76.1%, by gastrointestinal (GIS) biopsy in 11.7%, and by other biopsies in the remaining. Renal involvement was documented in 160 (98.2%) patients, while GIS involvement in 20.9%, heart in 13.5%, thyroid in 3.7% and bone marrow in 3.1%. In FMF patients, most common MEFV mutation was M694V (77.7%); and 66.7% of the patients had homozygous, 14.6% had compound heterozygous, and 18.7% heterozygous exon 10 variants. Mean age at diagnosis of amyloidosis was earlier in homozygotes (29.1) and compound heterozygotes (32.3) compared to heterozygotes (43.9) ($p = 0.001$). There was no difference in treatment responses, organ involvement, progression to end stage renal disease (ESRD) and mortality between monoallelic and biallelic exon 10 mutations ($p = 0.42$). While 44.3% ($n = 70$) of patients had chronic renal disease (CRD) at time of diagnosis, ESRD developed in 45.3% ($n = 73$) of patients. During follow-up, 55 patients underwent renal transplantation and recurrence of renal amyloidosis occurred in 24% of them. Mean creatinine and proteinuria levels at time of diagnosis were higher in patients with ESRD than those without ESRD ($p < 0.001$, $p = 0.03$ respectively). Progression to ESRD was significantly higher in patients with $GFR \leq 60$ ml/min at time of admission (%14.5 vs %41.7, $p = 0.005$, Figure 1). A total of 113 (70.2%) patients used biological agents, most commonly used biological agent was anakinra ($n = 81$). Canakinumab was used in 17 and other biological agents in 17 patients. Complete response was observed in 49.1%, partial response was observed in 6.2%, and progressive progression was observed in 21.7%. GIS and cardiac involvements were associated with progressive course ($p < 0.001$) and increased mortality ($p = 0.002$, $p < 0.001$, respectively), and overall mortality rate was 8.7%.

Figure 1 : Survival graphic of AA amyloidosis patients who developed ESRD according to their baseline GFR status

Conclusion: Increased rate of ESRD and progression of amyloidosis findings in patients who presented with $GFR < 60$ ml/min emphasize the importance of early diagnosis. Although mortality rate is very high in patients with AA amyloidosis due to FMF disease, it may be possible to reduce mortality with an effective treatment.

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2020-eular.3805

OP0275

REAL-WORLD CLINICAL BURDEN AND GLUCOCORTICOID USE IN PATIENTS WITH GIANT CELL ARTERITIS

R. Puneekar¹, P. Lafontaine², J. H. Stone³. ¹Sanofi, Cambridge, United States of America; ²Sanofi, Bridgewater, United States of America; ³Harvard Medical School, Boston, United States of America

Background: Giant cell arteritis (GCA) is a rare form of vasculitis usually manifesting in people aged ≥ 50 yr and is more common in women. Symptoms include headache, jaw claudication, fatigue, polymyalgia; and blindness if untreated. While risks of complications can be reduced with promptly administered high-dose glucocorticoids (GC; 20-60 mg for 2-4 wk, then slowly tapered), further risks of high GC exposure and related complications over the course of therapy remain.

Objectives: To compare GC use and GC-related complications in GCA patients (pts) vs a general population (GnP) cohort.

Methods: This retrospective, observational cohort study was based on Optum's de-identified Clinformatics® Data Mart Database (01/01/06-30/06/18, study period). The GCA cohort included pts with ≥ 1 inpatient or ≥ 2 outpatient claims ≥ 30 days apart with GCA-related diagnosis codes (ICD-9: 446.5x/ICD-10: M31.6x) between 01/01/06-30/06/17 (pt identification period) during which first occurrence of a GCA-related medical claim was set as index date (ID). The GnP cohort included pts without any medical claims for rheumatoid arthritis, GCA or polymyalgia rheumatica diagnosis codes during the study period, with their ID set as 12 mo from start of continuous health plan enrollment. Pts in both cohorts were required to be age ≥ 50 yr (on the ID) with continuous health plan enrollment ≥ 12 mo pre- and post-ID. Cohorts were 1:1 propensity score matched. GC use and incidence of GC-related complications were assessed from GC initiation, starting from the baseline period (12-mo pre-ID) to the end of GC use during the post-index period (ie the end of data availability, end of the study period, or death, whichever occurred first). Descriptive analyses included mean, standard deviation (SD) and median values for continuous variables, and frequency (n and %) for categorical variables. Continuous variables were compared between matched cohorts using t -tests and Wilcoxon sum rank tests. Categorical variables were compared between matched cohorts using Chi-square tests or Fisher's exact tests. Duration of GC use was analyzed using the Kaplan-Meier method and compared between matched cohorts using log-rank tests.

Results: There were 6071 pts included in each of the GCA and matched GnP cohorts; median age per cohort was 76 yr, median Elixhauser comorbidity index score was 3.0, and the majority (~75%) were women. The median follow-up duration was 44 and 48 mo in the GCA and GnP cohorts, respectively. A higher proportion of pts in the GCA cohort than the GnP cohort (90.6 vs 63.8%; $p < 0.001$) used GC. The mean (SD) duration of GC therapy was 230.5 (± 326.8) days in