Methods: A longitudinal observational study was performed (NEREA registry). Patients diagnosed with IPAF according to Fischer’s criteria were included from 2007 to 2019. The study was carried out by a multidisciplinary team (pneumologists, rheumatologists) in seven Hospitals of Madrid. The relative functional respiratory impairment, defined as a ≥ 5% decrease in the predicted forced vital capacity (FVC%) compared to the previous visit was set as the main outcome. Respiratory function was measured at baseline and every 6-12 months. Covariates included: a) sociodemographic, b) clinical, c) radiological pattern (non-specific interstitial pneumonia [NSIP]; usual interstitial pneumonia [UIP]; others); d) FVC% DLCO%; e) laboratory tests; f) therapy used. Survival techniques were used to estimate the incidence rate (IR) of relative functional respiratory impairment, expressed per 100 patient-year, for each specific confidence interval [95% CI].

Results: 79 IPAF were included, with a follow up of 462.8 patients-semeister and a maximum follow-up of 12.3 years. 79% were women with a mean age of 66±11 years. Along with obesity (40%), the most frequent comorbidities at baseline were hypertension, hypercholesterolemia, followed by ischemic heart disease. Baseline FVC% and DLCO% were 86.5±22.7 and 64.2±19.3, respectively. Distribution of IPAF classification criteria was: a) clinical domain: arthritis (46.2%); Raynaud’s phenomenon (35.6%) and mechanic hands (9.3%); b) serological domain: 80.8% positive ANA at >1/320 titer; 29% RF (> 40 IU/ml); 25% positive anti-Ro; c) morphological domain: 46.8% of NSIP and 36.7% of UIP. During the study period, 77.2% of patients (n=61) received treatment: glucocorticosteroids (n=52), mycophenolate (n=25), azathioprine (n=21), rituximab (n=15) and antibiotics (n=11). During the follow-up, 50 patients presented 111 relative functional respiratory impairment events over time. The estimated IR was 23.9 [19.9-28.1] per patient-year for patients with UIP, and 25% of the patients developed functional respiratory impairment at 16 months from diagnosis. IR was similar between patient gender, baseline overall comorbidity, baseline pulmonary functional tests, and age strata, with slight difference in patients >80 years of age. Patients with baseline associated emphysema (IR: 17.8 [10-31]) or without baseline associated fibrosis (IR: 21.1 [15-26]) had lower IR compared to the opposite (IR without emphysema: 24.5 [19.3-31]; IR with fibrosis emphysema: 27.3 [19.8-37.3]). As expected, IR was higher in UIP (32.2 [24-42]) compared to NSIP or any other pattern. With respect to serologic markers, patients with ANA titers ≥1/320 had a higher IR (26.7 [21-33]) in comparison with those with lower or non-titers of ANA (IR: 15.7 [9.9-25.1]).

Conclusion: In a multicenter registry of Madrid, we have performed a descriptive longitudinal study. IPAF were mostly women in their sixties. The most frequent clinical criteria were arthritis and Raynaud’s phenomenon. An NSIP radiological pattern predominated. At onset, patients have a slightly diminished lung function. The incidence rate of functional deterioration was estimated in 23.9% patient-year for patients with UIP. 50% of the patients developed pulmonary functional deterioration at 16 months from ILD diagnosis. This incidence rate was higher in patients with an UIP pattern, baseline fibrosis or ANA at medium-high titers.

Disclosure of Interests: None declared.


POS1428 IDENTIFICATION OF A WARM AUTOIMMUNE HEMOLYTIC ANEMIA (WAHA) POPULATION USING PREDICTIVE ANALYTICS OF A KNOWN CLINICALLY PROFILED COHORT


Methods: To identify a WAHA cohort using a collection of diagnostic codes, 2) bisected severe versus non-severe WAHA patients, 3) compare the frequency of comorbidities, anemia symptoms, treatments, diagnostic tests, and healthcare provider visits in these two groups, and 4) use a predictive model to validate clinical variables and prevalence estimates.

Background: WAHA is a rare disorder characterized by the destruction of red blood cells (RBCs) by pathogenic IgG autoantibodies, which has not been well studied. At the time this claims database analysis was carried out a diagnostic code specific for WAHA did not exist, which presented challenges in identifying patients with WAHA

Objectives: Despite the absence of a diagnostic code specific for WAHA we aimed to 1) identify a WAHA cohort using a collection of diagnostic codes, 2) bisect severe versus non-severe WAHA patients, 3) compare the frequency of comorbidities, anemia symptoms, treatments, diagnostic tests, and healthcare provider visits in these two groups, and 4) use a predictive model to validate clinical variables and prevalence estimates.

Methods: A de-identified longitudinal, patient-level claims database of 300 million US patients was used for this study. Patients with WAHA were identified based on a diagnosis code of “autoimmune hemolytic anemia” (AHA), chronic use of steroids (≥30 days) in the last 36 months, and chronic use of non-steroidal immunosuppressants. Patients were classified as severe if claims related to transfusion, high frequency of blood testing, high frequency interactions with a hematologist, and/or ≥2 ER visits per year were observed in the 36-month period. Codes for anemia symptoms, comorbidities, treatments, and diagnostic tests were grouped and analyzed within the most recent 12 months for each patient. Prevalence was estimated using Artificial Intelligence/Machine Learning (AI/ML) lookalike modeling, using known patients with WAHA as the positive training class.

Results: A cohort of 1,548 patients with WAHA was identified (n=631 were classified as severe while n=917 as non-severe). Median patient age was >65 years, and patients were evenly distributed by gender. The rate of disease-relevant claims was disproportionately higher in the severe cohort versus the non-severe cohort. Over the 12-month study period, there was a 61% higher rate for anemia symptomatology codes and a 570% higher rate for WAHA specific testing and monitoring codes in the severe cohort. Primary hypertension, hyperlipidemia, gastro-esophageal reflux, and evidence of chemotherapy use were also present in WAHA patients. All these conditions were observed more frequently in severe patients with the exception of lupus. Almost 44% of WAHA claims for the full cohort were associated with Hospital/Emergency care - 48% for the severe group. AI/ML modeling predicted patients using claims variables for hematologic anemia, other blood count abnormalities, and medical procedure claims commonly used for the diagnosis and management of WAHA. The predicted population supports reported US prevalence estimates of 30,000-49,000 patients.

Conclusion: We developed and validated a method for defining WAHA patients using de-identified claims data based on AHA ICD-10 codes and relevant treatments. We observed that while disease manifestations are generally the same in the severe and non-severe WAHA cohorts, there is an increased rate of occurrence in the severe cohort, which is not reproduced in any other pattern. As expected, IR was also associated with higher utilization of healthcare resources. The comorbidity of lupus was more commonly associated with non-severe WAHA patients. This may indicate that patients with a known diagnosis (in this case lupus) who are more closely monitored are less likely to reach the level of severity that would categorize them as patients with severe WAHA.


POS1429 IMPROVING ACCURACY OF SELF-REPORTED DIAGNOSES OF POLYMYALGIA RHHEUMATICA AND GIANT CELL ARTERITIS IN THE FRENCH PROSPECTIVE E3N COHORT: A VALIDATION STUDY

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Background: Giant cell arteritis (GCA) and polymyalgia rheumatica (PMR) are two associated inflammatory diseases that probably share common pathophysiological mechanisms. Data on environmental risk factors are lacking. Population-based cohort studies are the most adequate and less biased sources for identifying such factors. But case validation of disease diagnoses is the first necessary step for running such studies, even though it is not easy to perform.

Objectives: To assess the accuracy of self-reported GCA/PMR diagnoses and to develop algorithms to ascertain GCA/PMR in a large French population-based cohort, using combined data of a dedicated questionnaire and medication reimbursement database.

Methods: The E3N cohort study (Etude Epidemiologique auprès des femmes de la Mutuelle générale de l’Éducation Nationale) includes 96,995 healthy women born between 1949 and 1959, French women born between 1925 and 1950, recruited in 1990 and was designed to investigate lifestyle and environmental factors associated with chronic diseases. Participants completed biennial mailed questionnaires to update their health-related information, lifestyle characteristics, and newly diagnosed diseases. Women who self-reported a diagnosis of GCA and/or PMR were sent a specific validation questionnaire designed to ascertain the diagnosis including clinical, biological, and therapeutic data, along with ACR 1990 classification criteria for GCA and 2012 classification criteria for PMR. We then devised algorithms based on self-reported answers and a medication reimbursement database, and evaluated their accuracy, comparing them with diagnoses obtained from medical chart review.
Results: Among the 98,995 participants, 1,392 women self-reported GCA/PMR. The specific questionnaire was sent to 1,143 (82.1%) of the eligible women (249 women could not be contacted because of death or withdrawn consent) and response was obtained for 830 women (59.6%). Among them, 202 women provided sufficient medical data to ascertain a diagnosis and study accuracy of developed algorithms. 56 women were classified as ACG and 121 as PMR. Self-reported diagnoses alone had an accuracy of 87.6% with medical chart review. If women additionally self-reported a diagnosis confirmation by a physician and the use of glucocorticoids for ≥ 3 months, the accuracy was improved to 89.8%. For patients who did not respond to validation questionnaire, adding the use of glucocorticoids for ≥ 3 months in the reimbursement database also improved the diagnosis accuracy to 92.8%. These two designed algorithms also had the benefit of reducing the number of false positive cases by 10 and 16 respectively. Finally, 589 GCA and/or PMR cases were confirmed by our two devised algorithms: 401 cases with medical chart review and the cases detected by our algorithms in the cohort.

Conclusion: The accuracy of self-reported diagnosis of GCA/PMR was high in the E3N-cohort. Using additional data such as medication reimbursement and/or other self-reported data from a specific questionnaire, particularly the prolonged use of glucocorticoids led to a better accuracy with a very small number of false positive cases and seemed to be sufficient to correctly ascertain GCA and/or PMR diagnoses. With the validation of nearly 600 GCA and/or PMR cases in our cohort, we will be able to conduct epidemiological studies to identify risk factors of these diseases.