the number of emergency room visits, hospitalisation admissions and infections were investigated in the follow-up.

Results: One hundred twenty-three patients were included, 74 (60.2%) women, the mean age was 79.41 years old. Fifty-six (45.53%) received GCs alone (group 1) and 67 (54.48%) received GCs and MTX as an adjuvant treatment at some time during follow-up (group 2). 30 of these 83 patients (24.39% of total patients) received MTX in the first trimester after diagnosis (group 3). The cumulative doses of GCs, number of patients with relapses, visits to the emergency room and hospitalisation admissions are shown in the table 1. In none of these variables there were statistically significant differences among the three groups, except for the number of patients with relapses, which was greater in group 2 than in group 1 ($p=0.03$). The number of relapses in patients who received MTX early (group 3) was 56.7%, in the rest of patients (who only received GCs and those who started MTX after the first trimester) was 52.69%–33.3% of the patients in group 3% and 21.5% of the rest of the patients presented infections.

	Group 1: GCs alone	Grupo 2: GCs+MTX at some moment during the follow-up	Group 3: GCs and MTX since 1st trimester	P group 3 vs (1 +2)	P group 1 vs 2
Cumulative GCs doses at 6 month (mg±SD)	6193 ±6820	7028±11 230	5942,50±1827	0.629	0.646
Cumulative GCs doses at 12 months (mg ±SD)	7772 ±7154	8976±11 148	7987±2360	0.759	0.897
Cumulative GCs doses at 24 months (mg ±SD)	9340 ±7886	10814±11 150	9383±2522	0.608	0.903
N° of patients with relapses (35.71%)	20/56	42/67 (62.69%)	17/30 (56.7%)	0.430	0.003
N° of patients with emergency room visits	20 (35%)	22 (32.8%)	12 (24%)	0.508	0.849
N° of patients hospitalisation admissions	16 (28.57%)	21 (31.34%)	11 (36.7%)	0.370	0.844

Conclusions: Whilst MTX have been used in an effort to reduce toxicity from GCs and to improve efficacy of treatment our observational study shows that there is no benefit from adjunct MTX in GCA either in terms of efficacy or toxicity.

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Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.7148

THU0448 IS THERE AN ASSOCIATION BETWEEN ADULT IGA VASCULITIS AND CANCER?

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Background: An increased incidence rate of cancer has been reported in adult patients with IgA vasculitis (IgAV). These conclusions are mostly based on observations in severely ill, hospitalised subgroup of patients. Most of the studies allowed for a wide time interval between IgAV and cancer appearance, not necessarily reflecting a causative link.

Objectives: The aim of our study was to look for the potential association between IgAV and cancer in an unselected adult IgAV population.

Methods: We analysed medical records of prospectively followed, histologically proven adult IgAV cases at our secondary/tertiary rheumatology centre between 1 January 2010 and 30 June 2017, who were followed until 31 December 2017 and lived in a well-defined referral region. We identified cancer as concurrent with IgAV, if the patients had active cancer or a relapse of cancer or newly-diagnosed cancer diagnosed up to 6 months prior or 6 months after IgAV diagnosis. Cancers developing after 6 months of follow up were labelled as unrelated to IgAV. We used appropriate descriptive statistical methods, and the Fisher's exact and Mann-Whitney U tests to assess differences of clinical characteristics in acute phase of IgAV, between the cancer and non-cancer groups. The national prevalence and age adjusted incidence rates of cancer from a well-defined referral region were obtained from National cancer registry (NCR).

Results: During the 90 months of observation we identified 196 new IgAV cases. 2 patients died in the acute disease phase due to vasculitis, and 8 during the first 6 months of follow-up for reasons other than IgAV or cancer. 20 patients were lost to

follow-up. The remaining 166 patients (55% male, median (IQR) age 63^{43–76} years) were followed for a median (IQR) of 20^{11–32} months. At the time of IgAV diagnosis, 6/166 (3.6%) had active, previously diagnosed malignant disease (prostatic cancer in 4, among which one also had cancer of urinary bladder, and sarcoma in 2 patients). In 2/166 patients (1.2%) a new cancer was diagnosed at presentation and in 2/166 (1.2%) during follow-up. One of the patients with an active cancer of urinary bladder, was treated with antibiotics for urinary tract infection prior to IgAV diagnosis, and was also on chemotherapy. The patients with cancer were older (median age (IQR) 80^{77–83} vs. 64^{44–77} years; $p=0.002$) but their presenting features of IgAV, and the initial IgAV treatment did not significantly differ from those without cancer. At the end of the observation period, the prevalence of cancer in our cohort was 6.0%, compared to the 4.8% prevalence of cancer in our general population and the relative risk of cancer in our IgAV cohort was 1.25 (95% CI 0.687–2.29; $p=0.461$). The age adjusted incidence rate of cancer was 11.4 per 1000 patients per year. The annual age adjusted incidence rate of cancer in our citizens, acquired from NCR, was 8.9 per 1000 adults and the standardised incidence ratio for our IgAV cohort was 1.62 (CI 0.441–4.15; $p=0.472$).

Conclusions: In our cohort of unselected adult IgAV cases, we did not confirm the previous observations of the association of IgAV, and cancer.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.5359

THU0449 DIAGNOSTIC PERFORMANCE OF ANTINEUTROPHIL CYTOPLASMIC ANTIBODIES IN A COHORT OF UNSELECTED SPANISH PATIENTS

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Background: Antineutrophil cytoplasmic antibodies (ANCA) are the serological marker of some idiopathic systemic vasculitides, predominantly involving small and medium-sized blood vessels, such as granulomatosis with polyangiitis (GPA), microscopic polyangiitis (MPA) and eosinophilic granulomatosis with polyangiitis (EGPA), which are known as the ANCA-associated vasculitis (AAV). Nevertheless, ANCA have been reported in a number of other conditions.

Objectives: To retrospectively evaluate ANCA diagnostic accuracy in a cohort of unselected patients.

Methods: From January 2014 to December 2016 a total of 6781 serum samples with a test request for ANCA were submitted to the Immunology Department of a 1.000-bed tertiary teaching hospital from Barcelona (Spain), from both inpatients and outpatients.

Indirect immunofluorescence (IIF) was performed for all requests using a commercially available "Granulocyte Mosaic 13" (EUROIMMUN). IIF allowed recognition of three staining patterns: cytoplasmic (cANCA), perinuclear (pANCA) and atypical (xANCA). For the detection of antibodies against myeloperoxidase (MPO) and proteinase 3 (PR3) a chemiluminescent immuno-assay (CLIA) using commercially available "QUANTA Flash MPO/PR3" (INOVA diagnostics) was performed in patients with positive IIF.

We reviewed the clinical charts of patients that underwent ANCA testing and collected patients' diagnoses, as established by their treating physician one year after sampling. In the event of multiple ANCA test in a single patient we include only the first test request (we excluded 1323 tests performed in 661 patients). We also excluded 184 patients with insufficient information and 306 ANCA tests with no diagnostic purpose. Therefore the study population includes 4968 patients.

Statistical analysis was performed with Stata 14.2 (College Station, TX, USA). Diagnostic performance was assessed using sensitivity, specificity, positive likelihood ratio (LR+), positive and negative predictive values (PPV and NPV) and global efficiency. Confidence Intervals (CI) were calculated using Wilson method.

Results: Only 34 patients (0.68%) received a diagnosis of AAV: 25 MPA, 6 GPA and 3 EGPA.

	Sensitivity [CI 95%]	Specificity [CI 95%]	LR+ [CI 95%]	PPV [CI 95%]	NPV [CI 95%]	Efficiency [CI 95%]
Positive IIF	94.1% [80.9– 98.4]	87.6% [86.7– 88.5]	7.6 [6.8– 8.5]	5% [3.5– 6.9]	99.95% [99.83– 99.99]	87.7% [86.7– 88.5]
IIF Typical pattern	91.2% [77–97]	96.9% [96.3– 97.3]	29 [24.1– 35]	16.7% [12– 22.7]	99.94% [99.82– 99.98]	96.8% [96.3– 97.3]
cANCA/PR3 or pANCA/MPO	76.5% [60–87.6]	99.1% [98.8– 99.3]	83.8 [59.3– 118.4]	36.6% [26.4– 48.2]	99.84% [99.68– 99.92]	98.9% [98.6– 99.2]

The majority (87.1%) of patients had a negative ANCA test and only 12.9% were found positive by IIF. Among 643 positive patients IIF pattern distribution was: 457 (71.1%) atypical, 108 (16.8%) perinuclear and 78 (12.1%) cytoplasmic pattern.

Among patients with positive ANCA 32 (5%) had an AAV. Two patients with AAV had negative ANCA (one GPA and one EGPA).

Data of diagnostic accuracy of ANCA for AAV are showed in table 1:

Conclusions: ANCA testing with commonly commercially available methods has an excellent diagnostic performance for AAV in routine clinical practice, especially if a typical pattern is associated with proper antigenic specificity (cANCA with PR3 or pANCA with MPO).

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.3643

THU0450 DIAGNOSTIC RELEVANCE OF ORGAN BIOPSIES IN ANCA ASSOCIATED VASCULITIS

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Background: The diagnostic workup of ANCA-associated vasculitis (AAV) is a challenge due to the possible multi-organ involvement and the wide range of differential diagnosis. Before classification, vasculitis needs to be proofed by clinical or histopathologic signs.

Objectives: We aimed to evaluate specific histopathologic features of organ biopsies and their contribution to the diagnosis of vasculitis and to the classification of specific AAV subgroups.

Methods: Retrospective, single-centre cohort study in patients with granulomatosis with polyangiitis (GPA), microscopic polyangiitis (MPA) and eosinophilic granulomatosis with polyangiitis (EGPA), classified by ACR-criteria, who have received at least one organ biopsy. Characteristic histopathologic features were analysed and compared between AAV subgroups, organ systems and ANCA-status (Chi-square-test).

Results: 306 patients (GPA n=154, MPA n=58, EGPA n=94) diagnosed between 1990–2017 were included. All biopsies were taken at active stage of GPA, MPA or EGPA at initial diagnosis (n=415) or during flare of the disease (n=36). 168 patients had a renal biopsy, 185 patients had 283 non-renal biopsies (1 biopsy in 102 pts, 2 different organ biopsies in 68 pts, >2 in 15 pts). In kidney biopsies glomerulonephritis was described in 78.6%, unspecific inflammation in 26.8% and normal tissue in 1.2%. In non-renal biopsies vasculitis, granuloma, tissue eosinophilia, unspecific inflammation or normal tissue were reported in patients with GPA 32.9/29.4/21.2/71.8/9.4%, MPA 27.3/9.1/27.3/90.9/9.1% and EGPA 20.2/10.1/67.4/73.0/20.2%; p<0.0001 (lung n=103, 13.6/13.6/37.9/71.8/3.9%; skin n=35, 42.8/11.4/40.0/51.4/1.4%; upper respiratory tract n=98, 19.3/16.3/37.7/72.4/8.2%; peripheral nerves n=7, 14.3/14.3/0/14.3/42.8%; p<0.0001). According to the ANCA status the distribution was 31.1/25.2/28.3/68.9/10.7% in ANCA+patients and 20.8/11.1/62.5/79.2/19.4% in ANCA- patients (p<0.0001). Diagnosis of vasculitis was based on biopsy result in 2% of GPA, none of MPA and 8% of EGPA patients, but ACR criteria were only fulfilled including a characteristic biopsy in 6.6% of GPA and 35.5% of EGPA patients. Assignment to GPA, MPA and EGPA by application of the EMA algorithm were only possible in consideration of a characteristic non-renal biopsy in 2.8/0/21.9% and of a characteristic non-renal and/or renal biopsy in 4.7/14.6/28.1% of all patients.

Conclusions: Histologic proof of vasculitis remains the gold standard of AAV diagnostic, however the diagnostic value is most prominent for renal biopsies. Distribution of various histopathologic features is different among AAV subgroups (GPA, MPA, EGPA) and varies between different organ biopsies. While classification of GPA and MPA is only in a few cases based on histopathology, in EGPA a characteristic histopathology is necessary for classification in almost one third of patients.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.4177

THU0451 CAUSE OF DEATH IN PATIENTS DIAGNOSED WITH GIANT CELL ARTERITIS IN WESTERN NORWAY 1972–2012

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Background: Giant cell arteritis (GCA) is the most common systemic vasculitis in adults.

Objectives: To determine the causes of death in GCA patients during a 41 year period.

Methods: Hospital-based retrospective cohort study including patients diagnosed with GCA in Bergen Health Area during 1972–2012. Patients were

identified through computerised hospital records using the International Classification of Diseases (ICD)-coding system. Clinical information was extracted from patients' medical journals. We excluded patients if data were unavailable, if the reviewing rheumatologist found GCA to be an implausible diagnosis or if the American College of Rheumatology (ACR) 1990 classification criteria for GCA were not fulfilled. Information on cause of death was obtained from the Norwegian Cause of Death Registry (NCoDR). We grouped causes of death according to the European Shortlist for Causes of Death 2012-version (COD-SL-2012). Statistics Norway (www.ssb.no) provided background population data (all deaths during 1972–2012 in the Norwegian population ≥50 years of age). Statistical comparison was performed using mid-p values.

Results: The patient inclusion process and patient characteristics have been published previously.¹ A total of 792 patients were included, 566 (71.5%) females (mean age 73.5 years, SD 8) and 226 (28.5%) males (mean age 72.1, SD 9). 432 patients (54.5%) died during the study period (1 January 1972 – 31 December 2012). NCoDR had data on the cause of death for 431 of these. During the study period (1972–2012) there were 1635979 registered deaths in the general Norwegian population aged ≥50 years. The most frequent underlying causes of death in the overall study population were diseases of the circulatory system (53.4%), cancer (11.6%) and diseases of the respiratory system (10.0%). The most frequent causes of death in the general population were also diseases of the circulatory system (46.0%), cancer (22.8%) and diseases of the respiratory system (10.0%). The distribution of causes of death differed significantly between GCA-patients and the general Norwegian population (mid-p-value<0.001 both overall and stratified by sex). Absolute numbers of deaths and corresponding percentages of all causes of death stratified by sex are presented in figure 1.

Conclusions: The most frequent causes of death in our cohort of GCA-patients were diseases of the circulatory system, cancer and diseases of the respiratory system (including influenza and pneumonia). These were also the most frequent causes of death in the general Norwegian population aged ≥50 years, but the distribution of death causes differed significantly between GCA-patients and the general population. However, this might reflect differences in the composition of the populations that we were not able to adjust for. We aim to analyse this further by comparing our GCA-cohort with randomly selected age-, sex- and geographically matched control subjects.

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Disclosure of Interest: L. Brekke Grant/research support from: MSD, A. Diamantopoulos: None declared, B.-T. Fevang Consultant for: Lilly, Novartis, AbbVie, J. Assmus: None declared, C. Gjesdal: None declared

DOI: 10.1136/annrheumdis-2018-eular.3034

THU0452 ANTI-INTERLEUKIN-6 (TOCILIZUMAB) EXPERIENCE IN TAKAYASU'S ARTERITIS PATIENTS

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Background: Targeted therapies such as tumour necrosis factor inhibitors (TNFi) and anti-interleukin 6 (anti-IL-6) are increasingly being used in Takayasu's Arteritis (TA) patients who are unresponsive to corticosteroids±conventional immunosuppressives.

Objectives: The aim of this study was to evaluate the indications and efficacy of anti-IL-6 (tocilizumab) therapy in a case series of TA patients.

Methods: In the prospective database of the Hacettepe University Vasculitis Centre (HUVAM), 105 TA patients meeting the 1990 modified American College of Rheumatology (ACR) criteria were registered at the end of July 2017. Total 28