Introduction. – This study describes characteristics and outcomes of AAV patients included in a nationwide initiative collecting clinical and laboratory data on AAV since 2009.

Methods. – Sixteen vasculitis centres (9 x Nephrology, 4 x Rheumatology, 2 x Immunology, 1 x Pediatrics) have participated in web-based data collection, consisting of retrospective data supplied at entry and prospective follow-up with a visit recorded every 3–6 months. Statistical analysis included the Kaplan-Meier method and log-rank test for survival analysis.

Results. – A total of 569 patients (M/F 276/293, median age at diagnosis 58, range 12–88 years) were enrolled, 311 (55%) c/PR3-ANCA positive, 226 p/MPO-ANCA (40%) and 16 (3%) ANCA-negative. The mean time from diagnosis was 67 months (range 0–463). GPA was the most common diagnosis with 59%, followed by MPA (including renal-limited form) recorded in 35% and CSS in 4%. Cumulative organ involvement involved kidney in 89% (confirmed with renal biopsy in 75% of them), lungs in 61% and ENF in 39%. The estimated 5-year survival was higher in patients aged ≤ 65 years than in the older ones (85.3% vs. 67.2%, P < 0.001), in c/PR3-ANCA positive patients compared to p/MPO-ANCA (85.7% vs. 72.5%, P = 0.002) and in patients without severe renal vasculitis at diagnosis compared to those with ≤ 500 μL/L (86.5% vs. 66.8%, P < 0.001). Last available Vasculitis Damage Index (VDI) ranged between 0 and 16 (median 4). c/PR3-ANCA-positive patients were diagnosed younger (mean 52 vs. 60 years, P < 0.01), with better S-creatinine levels (mean 183 vs 257 μL/L, P < 0.01) and suffered from more relapses during follow-up (1.23 vs. 0.7/person, 47% vs. 34% with a relapse, P < 0.01) compared to p/MPO-ANCA positive.

Conclusion. – In the Czech population, GPA is the most common AAV, reflecting the northern European type. Older age and severe renal involvement are associated with higher mortality. Long-term survival is associated with relatively frequent relapses and significant morbidity in a number of patients.

http://dx.doi.org/10.1016/j.lpm.2013.02.034

A34
Infection risk in ANCA-associated vasculitis
M. Morgan1, A. Richter1, J. Flint1, M. Drayson1, C. Yiannakis1, D. Goldblatt2, L. Harper1
1. University of Birmingham, Birmingham, United Kingdom
2. UCL Institute of Child Health, London, United Kingdom

Introduction. – Infection is a frequent complication in anti-neutrophil cytoplasm antibody-associated vasculitis and is associated with increased morbidity and mortality. We investigated infection rates, infection risk factors and vaccine responses in a cohort of patients under long-term follow.

Patients. – Clinical data was collected retrospectively from patient records. Patients received prevnar 7, Men A,C,W135,Y and menitorix vaccines. Functional antibody titres were measured at baseline, 4 and 16 weeks and 2 years; anti-pneumococcal antibody (anti-PN) opsonophagocytic assay (OPA) was done at 16 weeks. All participants gave informed consent.

Results. – In 89 patients with median 5 (2–22) years follow-up overall rates were 1.5 infections/year and 0.9 infections requiring hospital admission/year. Serum immunoglobulin G (IgG) < 5 g/L was associated with infection. Ninety-two patients in established remission received vaccination with 16 weeks follow-up. Sixty-four patients were followed for 2 years. IgG < 6 g/L at vaccination was associated with low baseline anti-pneumococcal antibodies (anti-PN), CD19 B cell counts and CD4 T cell counts, increased age and continued immunosuppression. Functional antibody titres improved significantly from baseline following vaccination in most patients. Factors associated with poor responses (antibody titre post-vaccination < 0.35 u/mL for pneumococcus, < 0.1 u/mL for meningococcus and haemophilus) were baseline IgG < 6 g/L (P = 0.017) and continued immunosuppression. Patients who achieved anti-PN titres > 0.35 by 16 weeks generally persisted at 2 years (57–100% of patients). OPA showed a significant positive correlation with most anti-PN antibody titres tested. No increase in relapse was seen post-vaccination.

Discussion. – Patients with secondary immunodeficiency are at high-risk of infection and do not mount adequate vaccine responses. Strategies