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 (9x Nephrology, 4x Rheumatology, 2x Immunology, 1x 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 ENT 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 S-creatinine ≥500 μmol/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 μmol/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.

A34
**Infection risk in ANCA-associated vasculitis**

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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, 8 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

A33
**Premature ovarian failure in women with ANCA-associated vasculitis**

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Introduction – One of the serious side-effects of cyclophosphamide (CYC) is premature ovarian failure (POF). Data are scarce on the incidence of POF in women diagnosed with AAV treated with orally administered CYC.

Methods – We retrospectively studied the incidence of POF in all women diagnosed with active AAV before the age of 40 in our centre between 1980 and 2011. POF was defined as loss of ovarian function before the age of 40 years. This was retrospectively diagnosed as the time of the last menstruation followed by one year of amenorrhea.

Results – Forty-nine women aged under 40 years at diagnosis were identified. Twenty-seven patients were treated with CYC and 22 patients were treated with other immunosuppressive medication (n = 16) or co-trimoxazole. Age at diagnosis in CYC treated patients was 28 years (SD 8) and in patients not treated with CYC 27 years (SD 6). None of the 22 patients treated with other (immunosuppressive) medication developed POF; however ten patients are still at risk. POF developed in 14 CYC treated patients, five did not develop POF and eight women are still at risk (< 40 years). POF developed after a median period of 65 (IQR 8–89) months after the start of CYC. The mean age at POF was 36.3 years (SD 4.1). The median cumulative CYC dose in patients before the development of POF was 28.7 g (IQR 17.4–55.1) (n = 14) and at the age of 40 years in patients without POF, this was 14.5 (IQR 8.6–46.6) (n = 5), this was not statistically significant. Two of 14 patients who eventually developed POF had three children after start of CYC and three of five patients who did not develop POF after treatment with CYC had six children after start of CYC. Two of 12 patients not treated with CYC had four children after start of therapy. Five of the 14 patients were involuntary childless due to the development of POF.

Conclusion – POF frequently develops after oral cyclophosphamide therapy in young women with AAV. This study emphasizes the importance of development and use of alternative therapies in this group of patients.

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