Title:
Clinical and epidemiological studies on ANCA-associated vasculitis

Background
ANCA-associated vasculitides (AAV) are inflammatory diseases of unknown etiology, associated with anti-neutrophil cytoplasmic antibodies (ANCA). AAV are rare conditions often with multiorgan involvement. Before the introduction of immunosuppressive treatment, AAV was associated with high mortality. Modern treatment has improved survival substantially but patients are prone to disease- or treatment induced comorbidities. In this project we address a number of epidemiologic and outcome questions.

Methodology
Study 1: Incidence and predictors of severe infections in AAV: We performed a cohort-study of 325 AAV patients from a defined geographical in southern Sweden. By case record review, all events of severe infection (requiring hospitalization and intravenous antimicrobial treatment) were identified. Incidence rate of severe infections was estimated and possible predictors of severe infection were studied using uni- and multivariable cox-regression analysis.

Study 2: Impact of exposure to infections prior to onset of AAV: A study combining a case control and a cohort part, the case-control study examines prior infections as exposure and AAV as an outcome in 270 AAV patients and 2687 controls matched for sex, age and place of residence. Odds ratio (OR) between prior infection and later AAV development was calculated using a conditional regression model. The cohort study describes clinical characteristics and outcome of AAV with preceding infections.

Study 3: An update on epidemiology of AAV: we are updating the incidence and prevalence of AAV including cases diagnosed between 1997 and 2019

Study 4: Occurrence of interstitial lung diseases in patients with positive ANCA serology.

Preliminary results and conclusions

Study 1: Severe infection occurs in up to 40% of AAV patients and is associated with worse patients survival and increased organ damage. Old age and high disease activity independently predict severe infection. Severe infections are a common problem in AAV, leading to more extensive permanent organ damage and increased mortality.

Study 2: Prior infections, specifically respiratory and ear-nose-throat infections, are associated with later development of AAV. This could be observed for MPO (myeloperoxidase)-ANCA positive patients but not PR3 (proteinase-3)-positives. In certain individuals Infections might trigger the development of AAV. Vasculitis in these patients is more likely to be MPO-positive.