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Objectives: To evaluate the worldwide incidence and prevalence of ANCA vasculitis through a systematic review of the literature and meta-analysis.

Methods: A systematic search of MEDLINE and EMBASE search engines was carried out for studies that analyzed the incidence and prevalence of ANCA vasculitis in different geographical areas. Inclusion criteria: patients diagnosed with ANCA vasculitis according to ACR criteria/ Chapel Hill Consensus and adult patients (> 16 years). All ANCA vasculitis (microscopic polyangiitis, granulomatosis with polyangiitis, eosinophilic granulomatosis with polyangiitis) were considered. Exclusion criteria: editorials, conference abstracts, case or cases series reports and narrative reviews; insufficient description of the methods; lack of data to compute incidence or prevalence; and duplicate studies. Variables: Main variable: the pooled prevalence measured by the number of prevalent cases per million / person-year (95% CI) and the pooled incidence measured as the number of incident cases per million / person-year (95% CI). Secondary variables: the prevalence and incidence of each vasculitis ANCA and according geographic area. A meta-analysis was undertaken to estimate the pooled incidence and the pooled prevalence per million / person-years. The 95% CI and I² for heterogeneity were calculated.

Results: Twenty four studies were included. The pooled incidence (95% CI) was 12.2 per million / person-year (8.4-16.5) and the pooled prevalence (95% CI) was 130 per million / person-year (67.5-213). The individual incidence for each vasculitis was: GPA (6.7), MPA (5.9) and EGPA (1.6). The individual prevalence for each vasculitis was: GPA (69.3), MPA (21.9) and EGPA (13.5).

In the analysis by continents, the pooled incidence for GPA vasculitis was higher in Europe (7.5), while the pooled incidence for MPA vasculitis was higher in America (6.9) and for EGPA vasculitis it was higher in Asia (1.8). The pooled prevalence for GPA and MPA vasculitis was higher in Europe (83.9, 24.4 respectively) than in America (14.2, 12.8 respectively).

Conclusion: The pooled incidence and the pooled prevalence are higher in the case of GPA vasculitis compared to the rest of ANCA vasculitis. In general there is a predominance of incidence and prevalence of all ANCA vasculitis in the northern hemisphere compared to the south.

Figure 1. The pooled incidence ANCA vasculitis.

Figure 2. The pooled prevalence ANCA vasculitis

Disclosure of Interests: None declared

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POS0247

INTERSTITIAL ANCA-ASSOCIATED VASCULITIS ASSOCIATES WITH SEVERE KIDNEY INJURY INDEPENDENT OF GLOMERULONEPHRITIS

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Background: Antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV) is a small vessel vasculitis affecting multiple organ systems, including the kidney. Small vessels in the kidney include small-sized arteries (interlobular artery, afferent and efferent arteriole), capillaries (glomerular and peritubular capillary) and venules.

Objectives: Although crescentic ANCA glomerulonephritis (GN) is a common histological finding reflecting glomerular small vessel vasculitis, it is reasonable that manifestation of AAV could also contribute to interstitial small vessel vasculitis. Therefore, we here aimed to expand our current knowledge focusing on interstitial vasculitis in ANCA GN by systematic histological scoring of vascular lesions analogous to Banff.

Methods: A total number of 49 kidney biopsies with confirmed renal involvement of AAV at the University Medical Center Göttingen were retrospectively included between 2015 till 2020. A renal pathologist (SH) evaluated all biopsies and was blinded to clinical data collection and analysis. A detailed methodological section is provided in the Supplementary material and methods section.

Results: Since previous studies established that crescentic ANCA GN associates with severe kidney injury and acute deterioration of kidney function in AAV, we first systematically scored interstitial vasculitis in association with requirement of renal replacement therapy (RRT). Among all active and chronic tubulointerstitial lesions analogous to the Banff scoring system, the only association between severe kidney injury requiring RRT was observed for interstitial vasculitis in AAV reflected by peritubular capillaritis (ptc, p=0.0002) and arteritis (v, p=0.0069), affecting 5/49 (10.2%) and 11/49 (22.4%) of renal biopsies, respectively. Since it is known that severe deterioration of kidney function also correlates with crescentic ANCA GN, we next directly compared glomerular and tubulointerstitial lesions. The fraction of normal glomeruli was inversely associated with interstitial fibrosis (ci), total (ti) and inflammation in IFTA (i-IFTA), whereas glomerular crescents were associated with interstitial inflammation (i), tubulitis (t) and total inflammation (ti). In contrast, global glomerular sclerosis associated with less interstitial inflammation (i) but correlated with interstitial fibrosis (ci) and tubular atrophy (ct), confirming established mechanism that chronic glomerular injury leads to tubular atrophy and interstitial fibrosis. Interestingly, no association between interstitial vasculitis (ptc and v correlating with severe kidney injury) and any glomerular lesion in ANCA GN (also correlating with severe kidney injury) was observed, thereby confirming that interstitial vasculitis contributes to severe kidney injury independent of ANCA GN. By contrast, short-term renal recovery from RRT was equal in both groups, suggesting a distinct association with acute decline of kidney function at disease onset.

Conclusion: Taken together, by using the Banff scoring system we here expand our current knowledge of renal interstitial lesions in AAV revealing peritubular capillaritis and arteritis as important histological alterations associated with severe kidney injury in a considerable subset of AAV. Furthermore, our findings that interstitial vasculitis did not correlate with crescentic ANCA GN implicate that the characteristics of each vasculitis manifestation are independent and could further improve our understanding of mechanisms contributing to renal injury. These observations suggest that interstitial vasculitis in AAV may also affect long-term prognosis requiring further investigation.

Disclosure of Interests: None declared

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POS0248

PSYCHOMETRIC PROPERTIES OF OUTCOME MEASUREMENT INSTRUMENTS FOR ANCA-ASSOCIATED VASCULITIS: A SYSTEMATIC LITERATURE REVIEW

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Background: The OMERACT Vasculitis Working Group has defined a Core Domain Set of outcome measures for ANCA-associated vasculitis (AAV). However, the psychometric properties of available outcome measurement instruments in AAV, an essential consideration when choosing among instruments, have not been summarized.

Objectives: To systematically review and summarize the psychometric properties of outcome measurement instruments used in AAV.