Methods. – All MPA patients attending the center from 1990 to 2012 were included in the study. Patients were split into two cohorts (with diagnosis from 1990 to 2002 and with diagnosis from 2003 to 2012) and analysed with respect to disease manifestations, treatment regimens and side effects and mortality.

Results. – One hundred and twenty-three patients fulfilled ACR and/or EMA criteria and were included in the study. The median follow-up period was 22 (range 6–180 months, 46 [0–180] for the old cohort and 13.5 (0–92) for the new cohort, P < 0.0001). 102 patients (83%) had generalized and 14 (11%) severe disease on admission. Most patients received cyclophosphamide (cyc) and glucocorticoids for remission induction (n = 101, 82%). Complete remission/response was induced in 99 patients (80%), 17 (14%) were refractory (no follow-up in 2 pts.). Relapse was common (41% of patients). Forty-two percent retained a GFR of < 50 ml/min and 30% had persistent peripheral neuropathy. The duration of induction and cumulative cyc were significantly curtailed without any differences in remission rates (old vs. new cohort, median months: 6 [3–14] vs. 5 [3–11]; P < 0.0001; median cyc dose: 16 [3–56] vs. 8 [3–35]; P = 0.0095; complete remission: 47% vs. 44%, P = 0.750, partial remission: 40% vs. 31%, P = 0.345). The time to diagnosis, organ involvements, damage rates and relapse rates did not differ significantly between the old and new cohort; there was 1 death in the old vs. 0 in the new cohort.

Discussion. – Time to diagnosis is still long. Patients present most frequently with organ-threatening generalized disease on admission. Cyc can be spared and reduced to a median of 8 g without reducing remission rates. Refractory disease, damage and relapse rates could not be reduced over time.

Conclusion. – MPA is still a life- and organ-threatening disease requiring intense immunosuppression and being associated with high rates of damage.

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Long-term outcome of 123 microscopic polyangiitis patients in a monocentric German cohort
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Introduction. – The aim of the study was to determine the course and outcome of Microscopic Polyangiitis (MPA) in a German vasculitis center over 20 years.

Methods. – One hundred and twenty-three patients fulfilled ACR and/or EMA criteria and were included in the study. The median follow-up period was 22 (range 6–180 months, 46 [0–180] for the old cohort and 13.5 (0–92) for the new cohort, P < 0.0001). 102 patients (83%) had generalized and 14 (11%) severe disease on admission. Most patients received cyclophosphamide (cyc) and glucocorticoids for remission induction (n = 101, 82%). Complete remission/response was induced in 99 patients (80%), 17 (14%) were refractory (no follow-up in 2 pts.). Relapse was common (41% of patients). Forty-two percent retained a GFR of < 50 ml/min and 30% had persistent peripheral neuropathy. The duration of induction and cumulative cyc were significantly curtailed without any differences in remission rates (old vs. new cohort, median months: 6 [3–14] vs. 5 [3–11]; P < 0.0001; median cyc dose: 16 [3–56] vs. 8 [3–35]; P = 0.0095; complete remission: 47% vs. 44%, P = 0.750, partial remission: 40% vs. 31%, P = 0.345). The time to diagnosis, organ involvements, damage rates and relapse rates did not differ significantly between the old and new cohort; there was 1 death in the old vs. 0 in the new cohort.

Discussion. – Time to diagnosis is still long. Patients present most frequently with organ-threatening generalized disease on admission. Cyc can be spared and reduced to a median of 8 g without reducing remission rates. Refractory disease, damage and relapse rates could not be reduced over time.

Conclusion. – MPA is still a life- and organ-threatening disease requiring intense immunosuppression and being associated with high rates of damage.

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Microscopic Polyangiitis

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The role of MPO-positive cells and MPO deposition in glomerular capillary injury in patients with various glomerulonephritides
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Introduction. – Myeloperoxidase anti-neutrophil cytoplasmic antibody MPO-ANCA-associated glomerulonephritis (GN) is characterized by pauci-immune necrotizing glomerulonephritis (NCGN). We have recently reported that in human MPO-ANCA-associated GN MPO exists along the glomerular capillary walls near infiltrated MPO-positive cells, suggesting that MPO released from neutrophils may directly cause capillary injury (Clin Nephrol, in press). Here we investigated a possible role of MPO in the pathogenesis of glomerular capillary injury in various types of GN including MPO-ANCA-associated GN.

Methods. – We analyzed 1102 kidney specimens obtained from two patients with anti-GBM GN, 11 patients with lupus nephritis, two patients with post-streptococcal acute GN (PSAGN) 12 patients with Henoch-Schönlein purpura nephritis (HSPN) and 13 patients with IgA nephropathy (IgA N) as well as 20 patients with MPO-ANCA-associated GN. Glomerular infiltration of MPO-positive cells and deposition of extracellular MPO and endothelial cell injury were analyzed for samples of renal biopsies. Co-localization of MPO and CD34 deposition was also examined by immunofluorescence staining.

Results. – Glomerular infiltration of MPO-positive cells and deposition of extracellular MPO on glomerular capillary walls were observed in any...