A56 Outcomes in patients with granulomatosis with polyangiitis (Wegener’s) treated with short vs. long-term maintenance therapy

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Introduction. Study aim.—Determine outcomes in patients (pts) with Granulomatosis with polyangiitis (GPA) treated with long-term (> 18 mo) vs. short-term (≤ 18 mo) maintenance therapy.

Methods.—Retrospective chart review of pts seen from 1992 to 2010. Inclusion criteria:
- 1990 ACR criteria for GPA;
- induction therapy with daily cyclophosphamide (CYC) or weekly methotrexate (MTX);
- remission achieved;
- maintenance therapy initiated immediately following discontinuation of induction therapy;
- maintenance therapy with either MTX or azathioprine (AZA);
- duration of remission ≥ 18 mo;
- chronic documentation of remission and relapse.

Results.—One hundred and fifty-seven pts (137 pts in long-term group). Mean follow-up 3.1 years. Induction therapy with CYC used for severe disease (78%) and MTX (22%) for mild to moderate disease. Mean doses when maintenance therapy was begun: prednisone (pred) 19 mg/d, MTX 16.5 mg/wk and AZA 112 mg/d. No differences between groups in regards to initial organ manifestations, pred dose at rem, maintenance drug used or pulse dose methylprednisolone at diagnosis. Long-term group showed a 29% reduction in hazard ratio for relapse (HR 0.71[95%CI 0.43, 1.18], P = 0.18). Treatment for > 36 mo showed 66% reduction in hazard ratio for relapse (HR 0.34 [95%CI0.15, 0.76], P = 0.008). When length of treatment was considered as a continuous factor, longer courses had an inverse relationship with the risk of relapse (HR0.77 [95%CI 0.65, 0.92], P = 0.003) even after adjustment for pred dose (HR0.58 [95%CI 0.4, 0.83]; P = 0.003). Overall relapse rate of 58% (91/157) with 53% (48/91) of relapses off maintenance therapy. Of pts on therapy at relapse 52% were on < 15 mg/wk MTX and 75% on ≤ 50 mg/d AZA. No differences between the two groups in overall adverse events or GPA related morbidity.

Discussion.—Pts receiving long-term maintenance therapy have fewer relapses and have a similar adverse event profile as pts treated for < 18 mos.

Conclusion.—Discontinuation and low doses of maintenance therapy are associated with a high relapse rate.

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A57 Characteristics and outcome of patients with granulomatosis with polyangiitis (Wegener’s) and microscopic polyangiitis on renal replacement therapy – Data from the ERA-EDTA registry

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Introduction.—This study describes the incidence and outcome of patients starting RRT for end-stage renal disease (ESRD) due to ANCA-associated vasculitis (AAV).

Methods.—Twelve renal registries, providing data to the ERA-EDTA Registry for at least 16 years between 1991 and 2010, participated. Survival analysis used the Kaplan-Meier method and Cox regression in GPA (granulomatosis with polyangiitis, Wegener’s) and MPA (microscopic polyangiitis) patients and in other RRT patients.

Results.—A total of 2371 AAV patients (1650 GPA and 721 MPA patients) were identified (1.21% of all 195,826 incident RRT patients), representing a crude incidence of 1.01 per million population (pmp) for GPA and 0.44 pmp for MPA. In the northern countries, the incidence of RRT for ESRD due to GPA was higher than MPA, while in the southern countries MPA prevailed. A higher percentage of GPA-patients recovered independent renal function within 90 days compared to all patients (6.7% vs. 1.5%, P < 0.0001). Three hundred and sixty with GPA (21.8%) and 139 with MPA (19.3%) received a kidney transplant. The 10-year survival probability on RRT was 31.5% (95% confidence interval 28.5–34.5%) in GPA and 24.2% (20.2–28.3%) in MPA. Patient survival on RRT, on dialysis and survival after kidney transplantation did not differ between AAV and non-AAV non-diabetic patients. Graft survival adjusted for age and sex was better in GPA-patients than in non-AAV patients without DM (hazard ratio 0.79 [95% confidence interval 0.66–0.95]). Patients with GPA and MPA were more likely to die from infection and less likely from cardiovascular events compared to all patients.

Conclusion.—Geographical differences in the incidence of RRT for ESRD due to GPA and MPA copied the previously described distribution of both diseases. The overall survival of AAV patients was similar to that of patients with other diagnoses. Our results suggest that patients with AAV, and particularly GPA, are suitable candidates for kidney transplantation with favorable graft survival outcomes.

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A58 Occupational and environmental risk factors in chronic periaortitis: A case-control study

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Introduction.—Chronic periaortitis (CP) is a rare condition characterized by the presence of a fibro-inflammatory retroperitoneal periaorti c and periilar tissue. CP is usually considered an idiopathic disease, but an