detection in the abdominal aorta ($P = 0.01$) as being significantly associated with MCVE. Plaque in the aorta was significantly ($\chi^2$ test) associated with high-risk status ($P < 0.001$), while BMI and high-risk status were independent variables ($P = 0.64$). Thus, a BMI $> 30$ kg/m$^2$ and/or a high-risk status were strongly associated with MCVE ($P = 0.003$).

Finally, IMT distinguished between patients with early or late MCVE. IMT was correlated with the time to MCVE ($r^2 = 0.78$, $P = 0.008$).

**Conclusion.**—These results suggest that factors associated with a higher MCVE risk in SNV patients are NCEP/ATP III-defined high-risk status and BMI $> 30$ kg/m$^2$. Carotid IMT measurement in SNV patients could help identify those at risk of early MCVE.

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


**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%CI 0.15, 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% ≤ 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 (ppm) for GPA and 0.44 ppm 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 received 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 retroperi toneal periaortic and peri iliac tissue. CP is usually considered an idiopathic disease, but an
association between asbestos exposure and risk of CP has been reported. The aim of this study was to investigate the role of occupational and environmental exposure to asbestos, metals, pesticides, silica, organic solvents in CP; we also evaluated the possible interaction with tobacco smoking.

Patients.– We enrolled 90 consecutive patients with CP diagnosed at or referred to our Department from all over Italy between 2004 and 2012: 270 subjects recruited from the general population and matched with the patients for age, sex and geographic origin served as controls. All the study subjects were asked to fill in a questionnaire in order to assess individual risk factors (such as cardiovascular disease, abdominal surgery, drug use and smoking) and occupational and environmental exposure to asbestos, organic solvents, metals, other industrial chemicals, and pesticides.

Results.– Asbestos exposure was over-represented among cases [OR 4.05 (2.28–7.19), P < 0.00001], exposure to other agents also tended to be more frequent in the patient group, but for none of them statistical significance was reached. Among cases, there was an excess prevalence of smokers [OR 3.16 (1.67–5.99), P = 0.0003]. The pack-year index was also significantly higher in the patient group [median (IQR) 31(20–44) vs. 20(9–37), P < 0.0001]. Interestingly, we found a positive interaction between asbestos exposure and smoking [OR 10.10 (4.28–23.84), P < 0.0000001].

Conclusion.– Exposure to asbestos may be a significant risk factor for the development of CP. In addition, tobacco smoking also increases the risk of CP. Asbestos and tobacco smoking significantly interact and, together, substantially increase the risk of developing the disease.

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A59
Behçet’s disease in Budd-Chiari Syndrome

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Introduction.– Behçet’s disease (BD) is a systemic vasculitis characterized by oral and genital ulcerations, ocular inflammation and venous thrombosis. BD is a well recognized cause of Budd-Chiari Syndrome (BCS). Data are lacking regarding the relationship between BD and BCS.

Patients.– Using a cohort of 111 patients with BCS, we investigated the relationship between BCS and BD in 19 patients with combined diseases and compared the results to 92 BCS patients in whom BD was excluded.

Results.– Median follow-up for the study group (n = 111) was of 53 months (range 1–141 months). BD patients with BCS were younger (P = 0.025), more frequently of male gender (P < 0.0001), originated more frequently from North Africa (P = 0.0002), had higher plasma C-reactive protein level (P < 0.0001) and presented more frequently with inferior vena cava (IVC) obstruction (P < 0.0001), as compared to BCS without BD (n = 92). Among BD patients with BCS, three patients were treated with recanalisation of the vena cava and of the hepatic veins, none patient had a TIPS and one had liver transplantation. Transjugular intrahepatic portosystemic shunt (TIPS) was significantly less performed in BCS patients with BD as compared to those without BD (P = 0.003). Eighty nine percent of BCS patients with BD received corticosteroids and/or immunosuppressive therapy. The 5-years survival rate was of 74% (CI 95% 55–100) and 79% (CI 95% 71–88) in BCS patients with and without BD (P = 0.47), respectively.

Conclusion.– This study shows that despite a higher frequency of additional IVC thrombosis, short-term prognosis of BCS patients with BD does not differ from BCS patients without BD. TIPS is less performed because of technical issues and spontaneous improvement with medical therapy. Medical treatment with anticoagulation and immunosuppressive agents seems to be efficient and sufficient to control BCS symptoms.

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A60
The unmet need in Behçet’s disease: Most patients are not in complete remission in the long-term follow-up

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Introduction.– Behçet’s disease (BD) is a systemic disorder with a remitting-relapsing course. As complete remission should be aimed in all inflammatory diseases, we investigated the frequency of complete remission in routine practice in BD.

Methods.– One hundred and thirty patients with BD (F/M: 67/63, mean age: 43.23 ± 11.7 years) classified according to ISG criteria were included, retrospectively. The data for active organ manifestations and treatment protocols were evaluated, both for the current visit and in the last month. Patients having at least one disease manifestations were categorized as active.

Results.– Totally, 857 visits were overviewed. Mean visit number was 6.5 ± 2.7 (1–10), mean follow-up duration was 53.54 ± 41.79 months.

Conclusion.– This study shows that despite a higher frequency of additional IVC thrombosis, short-term prognosis of BCS patients with BD does not differ from BCS patients without BD. TIPS is less performed because of technical issues and spontaneous improvement with medical therapy. Medical treatment with anticoagulation and immunosuppressive agents seems to be efficient and sufficient to control BCS symptoms.

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