of outcome. C-ANCA positive patients had better outcome than P-ANCA positive.

**Discussion.**—There was a significant difference in outcomes between patients in the focal and sclerotic group at 5 yrs. Previous validation studies [2] have differed with regard to outcomes in the crescentic and mixed groups, which in our study were similar. The greatest improvement in eGFR at 1 and 5 yrs was in the crescentic class. Percentage normal glomeruli, degree TA and starting eGFR were previously correlated with outcome and our study confirmed these findings.

**Conclusion.**—This large study provides further validation of the prognostic use of Berden’s classification of renal AAV, and in addition highlights the importance of proportion of normal glomeruli, TA, starting eGFR and ANCA type as prognostic factors.

**References**


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**A48**

**Diffuse alveolar haemorrhage (DAH) in levamisole-adultered cocaine abuser resolved by rFVIIa infusion**

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**Introduction.**—Vasculitis induced by levamisole-adultered cocaine is a new entity described since 2008. It mainly involves skin with necrotic erythematous purpuric lesions located at nose, cheeks, earlobes and extremities. Another main complication is agranulocytosis. Fever, malaise and arthralgia/myalgia are usually present. It is associated with presence of autoantibodies: ANA, ANCA with predominant anti-MPO specificity, antiphospholipids antibodies and low C3 values. Differently from ANCA-Associated vasculitis, kidneys and lung are rarely involved. We report a case with severe DAH in cocaine abuser. Neither steroid pulses nor plasma-exchange were able to stop lung bleeding, which only resolved after rFVIIa administration.

**Results.**—A 37-yo cocaine abuser male was admitted with 3 days history of malaise, thoracic pain, diffuse arthralgias. Physical examination revealed purpura and necrosis on lower extremities and earlobes, perforated septum with active crushing. CT scan showed right interstitial diffuse alveolar infiltrates, suggestive for DAH. Bronchoscopy confirmed DAH by identification of ongoing bleeding from the right bronchial tree; no positive bacterial, fungal or viral organisms were identified. Bronchial biopsy proved vasculitis. Pt rapidly developed haemoptysis and respiratory insufficiency. A chest X-ray demonstrated bilateral rapidly worsening air space disease. Urinalysis revealed ematuria and proteinuria, sCreatinine was normal. Laboratory exams showed ANCA anti-MPO at 67.9 RU/mL (nv 19.99), antiphospholipids antibodies were negative. IV antibiotics, high doses methylprednisolone (1 g iv), transfusion support and plasmapharesis were administered. On day 7 the bleeding was still ongoing, thus rFVIIa was given (7 mg i.v.), in order to obtain...
hemostatic effect. For uncontrolled hypoxia, he was transferred to the ICU and intubated. Pulmonary bleeding stopped after rFVIIa administration iv. Bronchoscopy on day 8 was negative for bleeding. He was depending on mechanical ventilation but the arterial oxygen pressure (pO2)/(FiO2) ratio increased the following day. Over the next 10 days, the situation improved and pt was discharged from the ICU.

No thromboembolic complications were observed after administration of rFVIIa i.v.

**Conclusion**—This case suggests that in levamisole-adultered cocaine induced syndrome, severe lung and renal involvement can occur and rFVIIa i.v. should be considered in severe DAH not responding to therapy.

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**A49**

**ANCA-associated vasculitis (AAV) and thrombotic microangiopathies (TMA): A retrospective histological study**

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**Introduction**—A number of case reports of vasculitis with associated TMA are progressively reported in the literature. Moreover studies in animal models suggest that complement activation is crucial in the pathogenesis of ANCA-associated vasculitis (AAV) [1], supplying a possible predisposing pabulum for TMA appearance. At our knowledge no data are available describing the possible association between vasculitis and TMA. We studied retrospectively our historical AAV cases to document the presence of histological signs of TMA.

**Patients**—We retrospectively examined a series of 39 consecutive patients diagnosed with renal biopsy as having AAV between 1990 and 2012. Patients had a mean follow-up of 86 months. Renal histology was reviewed by an independent pathologists, blinded for renal outcome.

**Results**—Ten out of thirty-nine patients died during follow-up. Six out of thirty-nine (15%) patients were identified as having clear signs of TMA. Four out of six patients of the TMA group had often an associated histological evidence of fibrinoid necrosis while the non TMA group presented an overall greater risk for renal failure on the basis of recently risk stratification scores in renal vasculitis; nonetheless the TMA group showed a significantly worse renal outcome (P < 0.05) (figure 1).

**Discussion**—Our preliminary data suggests that, similarly to other glomerulonephritis, TMA should be evaluated as an associated historical pattern in renal vasculitis. TMA seems to determine a worse renal prognosis; consequently recognition of TMA associated features may allow a more appropriate management (i.e. plasmapheresis) of acute renal failure in these cases.

**Reference**


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**A50**

**The relative risk of aortic aneurysm in patients with giant cell arteritis compared with the general population of the UK**

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**Introduction**—Giant cell arteritis (GCA) is the most common systemic vasculitis; small studies have demonstrated 22% of these patients will develop an aortic aneurysm [1]. The British Society of Rheumatology and the European League Against Rheumatism has highlighted the need for a precise estimate of the increased risk of aortic aneurysm to inform definitive advice regarding screening of patients with GCA [2,3].

**Patients**—This parallel cohort study includes 6999 patients with GCA from the UK General Practice Research Database (GPRD). Non-GCA patients were matched on a 6:1 ratio on: GP practice, year of birth and gender. A competing risk model using aortic aneurysm as the primary outcome and death as the competing risk was used to determine the relative risk (subhazard ratio) between non-GCA and GCA subjects, after adjustment for cardiovascular risk factors.

**Results**—Comparing the GCA cohort with the non-GCA cohort; after adjustment for BMI, smoking, alcohol, hyperlipidaemia, lipid lowering medication, hypertension, anti-hypertensives, diabetes, cardiovascular disease, cerebrovascular disease and peripheral vascular disease; the subhazard ratio for aortic aneurysm (95% CI) was 1.92 (1.52 to 2.41). Significant predictors of aortic aneurysm were as follows: being a smoker 3.37 (2.61 to 4.37), previous history of diabetes 0.32 (0.19 to 0.56) and previous history of cardiovascular disease 1.98 (1.50 to 2.63). In a multivariable model of the GCA cohort alone, male gender 2.10 (1.38 to 3.19), smoking 3.79 (2.20 to 6.53), and diabetes 0.19 (0.05 to 0.77) were significant predictors of aortic aneurysm.

**Discussion**—This is the largest cohort study of patients with GCA to date, and demonstrates a two-fold increased risk of aortic aneurysm. Other risk factors for aortic aneurysm, including male gender, age, and smoking, and a protective effect of diabetes, are however also important in GCA patients.

**Conclusion**—GCA should be considered within the context of other risk factors for aortic aneurysm, rather than needing separate screening.