(95% CI 1.18–24.20)), cumulative cyclophosphamide dose \( (HR 15.98 (0.005–0.83)) \), haemoglobin level at the end of follow-up \( (HR 0.6 (0.262–0.987)) \), serum PR3-ANCA levels at onset \( (HR 0.97 (0.995–0.99)) \) and history of prior CVE \( (HR 5.3 (1.015–27.69)) \). A cumulative RTX dose < 6 g was associated with higher maintenance prednisolone dose \( (P = 0.016) \).

Conclusion. – CVE/death risk in AAV patients is especially high within the first 1 and 5 years of diagnosis. Prior CVE, low serum PR3-ANCA levels at onset and lower haemoglobin at the end of follow-up are associated with increased cardiovascular risk. Intensive immunosuppressive treatment of AAV at onset and avoidance of high long-term prednisolone dosage may have a protective effect against atherosclerosis. Rituximab therapy was associated with a steroid-sparing effect.

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The value of a patient global assessment for disease activity in granulomatosis with polyangiitis


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Introduction. – There is increasing awareness of the importance of patient global assessment (PiGA) scores in clinical research for rheumatic diseases. The purpose of this study was:

– to describe the distribution of PiGA scores in patients with granulomatosis with polyangiitis (GPA, Wegener’s) enrolled in a clinical trial;
– explore discordance between PiGA and physician global scores for disease activity (PhGA);
– explore if PiGA scores during disease remission are associated with future disease relapse.

Methods. – Data from the Wegener’s Granulomatosis Etenacept Trial (WGET) were analyzed. PtGA and PhGA were assessed on visual analog scales (0–100). Disease activity was assessed with the Birmingham Vasculitis Activity Score for Wegener’s Granulomatosis (BVAS/WG) and remission defined as BVAS/WG = 0. Disease relapse was defined as BVAS/WG > 0 after remission was achieved. PtGA-PhGA discordance was defined as a difference between the two measures of ≥ 20. Mixed linear regression models were used in longitudinal analysis of PtGA scores.

Results. – The 180 subjects in WGET had 1719 study visits. Baseline mean PtGA was 64.2 (sd = 27.4) and mean PhGA was 55.5 (sd = 23.4). PtGA-PhGA discordance occurred in 54% of subjects at baseline. Having a new disease at baseline was inversely associated with PtGA-PhGA discordance \( (OR = 0.37, 95\% CI 0.20–0.68) \). PtGA-PhGA discordance was not associated with age, sex, renal disease, or pulmonary disease. Subjects were in disease remission during 62% of study visits. Mean PtGA scores were 4.52 points higher during visits immediately prior to relapse than at other remission visits \( (95\% CI 0.66–8.40, P = 0.03) \).

Conclusion. – PtGA-PhGA discordance is common in GPA. A rise in PtGA during times at which physicians defined the patient to be in remission is associated with subsequent relapse. These findings support the addition of PtGA as an outcome measure for GPA.

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Patient reported outcomes in ANCA-associated vasculitis. A prospective comparison between BVAS and RAPID3 on an MDHAQ

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Introduction. – ANCA-associated vasculitis (AAV) is a rare group of relapsing diseases comprising GPA, MPA, and EGPA. It is desirable to quantitate patient symptoms, but difficult for the staff to manage diagnosis-specific questionnaires. The Multi-Dimensional Health Assessment Questionnaire (MDHAQ) has been documented to be effective in many rheumatic diseases. Therefore, we compared a patient-only index termed the “routine assessment of patient index data 3” (RAPID3) on an MDHAQ to the Birmingham Vasculitis Activity Score (BVAS).

Patients. – Patients with AAV treated at Rush University from Jan 2010 to May 2012 completed MDHAQ for four consecutive visits approximately 6 months apart. An independent investigator scored RAPID3, which comprises three Core Data Set measures on the MDHAQ for function, pain, and patient global assessment (PATGL); scores range from 0 to 30, with higher scores being worse. BVAS was calculated at each visit and compared to RAPID3 and PATGL. Scores were compared using Spearman non-parametric correlations. Linear regression was used to adjust for other variables.

Results. – Twenty-nine patients with AAV consented, 22 had GPA, five MPA and two EGPA. The mean age was 54.1 years and 77% were females. The median BVAS was 5.5 (IQR: 2.7–9.2), median RAPID3 was 6.8 (IQR: 2.3–13.6), and median PATGL was 3.2 (IQR: 1.4–6). RAPID3 correlated with BVAS at each visit \( (\rho = 0.45, 0.75, 0.73, 0.54 \) with p values of 0.02, < 0.0001, 0.002, and 0.05 for visits 1 to 4, respectively) and PATGL correlated with BVAS at three out of four visits, independently of RAPID3 \( (\rho = 0.24, 0.75, 0.64, 0.59 \) with P values of 0.23, < 0.0001, 0.01, 0.01 for visits 1 to 4, respectively).

Conclusion. – RAPID3, a patient-only index, correlates significantly with the BVAS. RAPID3 can be calculated in 5 seconds and does not require physician input, laboratory or imaging information. PATGL may also reflect disease activity. In the face of increased expenses and busy practices, such instruments may help document patient status and add to clinical decisions.

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Severe fatigue and psychological morbidity in ANCA-associated vasculitis

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Introduction. – Patients with ANCA-associated vasculitis (AAV) report fatigue as a main debilitating symptom, but little systematic research has been done to assess its impact. We aimed to characterize the