Introduction. – Granulomatosis with polyangiitis (Wegener’s) (GPA) and microscopic polyangiitis (MPA) are multisystem diseases known collectively as the antineutrophil-cytoplasm antibody associated vasculitides (AAVs). Damage accrued in AAVs can be quantified by use of the Vasculitis Damage Index (VDI), a validated 64 item checklist divided into 11 categories which captures effects of both disease and therapy.

Methods. – Data were merged from a long-term follow up questionnaire completed for patients from four European Vasculitis Study Group (EUVAS) therapeutic trials (n = 535). Two hundred and seventy patients of the 535 patients from the trials had VDI at baseline, 6 months, 12 months and long-term follow-up. Treatment related items were defined a priori. Categorical variables are expressed as frequencies with percentages. Chi-squared or Fisher’s exact tests were used to compare groups in terms of change in VDI scores over time.

Results. – Time of follow up was 6.94 years (SD 2.04). The frequency of treatment-related cardiovascular VDI damage items rose significantly (most P < 0.001) over time (Supplementary data). By long-term follow-up, hypertension was recorded in 41.5%, angina/bypass in 8.1%, cerebrovascular events in 3.7% (P = 0.002), and myocardial infarction in 4.4% (P = 0.003). The frequency of corticosteroid related VDI damage items also rose significantly. At long-term follow-up, osteoporosis was recorded in 14.1%, diabetes in 10.4%, in 9.3%, and muscle atrophy and weakness in 7.4%. The frequency of malignancy and gonadal failure also rose significantly over time, with the percentages of affected patients at long-term follow-up being 12.6 and 4.1% (P = 0.021). 101 (37.4%) of the 270 patients had no treatment related damage at long-term follow up.

Discussion. – Cardiovascular disease, diabetes, osteoporosis and malignancies are significantly increased in patients with AAV.

Conclusion. – Long-term treatment related damage in AAV can be quantified using the Vasulitis Damage Index (VDI).

Supplementary data associated with this article can be found on the website of La Presse Médicale (http://www.em-consulte.com/revue/lpm).

Table I Frequency of recording of VDI items related to treatment over the course of long-term follow-up (n = 270).

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P151
Factors associated with long-term damage in the AANA-associated vasculitides: An analysis of cohorts from the European vasculitis study group (EUVAS) therapeutic trials

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Introduction. – Granulomatosis with polyangiitis (Wegener’s) (GPA) and microscopic polyangiitis (MPA) are antineutrophil-cytoplasm antibody associated vasculitides (AAVs). Damage accrued in AAVs can be quantified by use of the vasculitis damage index (VDI).

Methods. – A long-term follow-up questionnaire was completed for patients from the European Vasculitis Study Group (EUVAS) trials (n = 535). Two hundred and ninety-six patients had VDI and steroid use data available at long-term follow-up (LTFU). Multiple linear and logistic regression was used to look for independent associations between baseline measures (age, creatinine and BVAS scores), cumulative factors (number of relapses and duration of corticosteroids used) and VDI scores at LTFU.

Results. – Patients with higher damage scores at long-term follow-up were: those on steroids at the last visit (P = 0.003), those with more relapses (P = 0.027), who were older (P = 0.052), with higher baseline creatinine (P = 0.002) and BVAS scores (P = 0.041). Age at entry was significantly associated with osteoporosis [OR 1.046 per year (95% CI 1.02 to 1.08), P = 0.046], malignancy [OR 1.043 (95% CI 1.01 to 1.08), P = 0.011], and cerebrovascular accident [OR 1.062 (95% CI 1.01 to 1.12), P = 0.034]; Creatinine at entry was significantly associated with hypertension [OR 1.002 per mmol/dL (95% CI 1.001 to 1.003), P = 0.001], angina/bypass [OR 1.001 (95% CI 1.000 to 1.003), P = 0.022], and CVA [OR 1.002 (95% CI 1.00 to 1.003), P = 0.022]; duration of corticosteroids was significantly associated with hypertension [OR 1.03 per month (95% CI 1.013 to 1.046), P < 0.001], and cataract [OR 1.032 (95% CI 1.002 to 1.06), P = 0.036]; number of relapses was significantly associated with osteoporosis [OR 1.35 per relapse (95% CI 1.036 to 1.77), P = 0.026], and CVA [OR 1.74 per relapse (95% CI 1.16 to 2.61), P = 0.008].

Discussion. – Long-term damage in the AAVs is predicted by severity of initial disease, age, number of relapses and duration of steroids.

Conclusion. – Addressing these risk factors may reduce long-term damage in the AAVs.

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P152
Markers for work disability in ANCA-associated vasculitis


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Introduction. – ANCA-associated vasculitis (AAV) commonly affects those of working age. The traditional outcome of mortality has been transformed by immunotherapeutics. In consequence, the measurement of other, patient-centred, outcomes has become increasingly relevant. Few studies have examined work disability, an important outcome for both patient and society. We aimed to assess employment status in AAV patients and identify putative predictors of their work disability.

Methods. – A multi-centre, cross-sectional study was undertaken. AAV cases were recruited according to consecutive clinic attendance.

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Subjects completed a questionnaire, which determined employment status and, in addition, assessed psychosocial factors (depression, fatigue, sleep, pain, coping). Concurrently, disease factors were recorded in the clinic (including BVAS, VDI, ANCA, co-morbidity, drugs and clinical phenotype). From the data of those subjects of working age, a multivariable model was developed using forward stepwise logistic regression to identify the independent associations of work disability, defined by those subjects reporting unemployment secondary to their health. Results are expressed as odds ratios (OR) and 95% confidence intervals.

Results—Of the 410 participants (86% response rate), 149 (36.3%) were employed (86 full time, 49 part time, 10 house persons, four students), 197 (48.0%) retired and 54 (13.2%) unemployed secondary to their health. Of those of working age, 25.9% were considered work disabled. Fatigue (OR 7.1, 1.5–33.1), depression (OR 4.4, 1.8–10.8), VDI > 4 (OR 3.9, 1.01–14.7) and being overweight (OR 3.4, 1.3–8.9) were independently associated with their unemployment.

Conclusion—A quarter of work aged AAV subjects reported unemployment as a result of their health and are characterised by high levels of fatigue, depression, disease damage and being overweight. However, it is unclear whether all these factors predict unemployment, or are consequences of it. Longitudinal studies are required to determine their causality.

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P154
Long-term outcome of GPA and MPA in a population based cohort

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Introduction—The outcome of GPA and MPA has dramatically improved since the introduction of steroids and cyclophosphamide, especially for patients entered into clinical trials such as EUVAS, but these may not be representative of the general population (trials will exclude early fatal cases for example) There is still a lack of long-term data on outcome, in particular mortality. We have maintained a prospective population register (NORVASC) of all patients with GPA and MPA since 1988. The aim of this study was to report the long-term mortality of GPA and MPA from our unselected well-defined population and to compare the periods 1988–1999 and 2000–2010. Treatment followed standards of care for the period, cyclophosphamide was mainly given IV.

Patients—Patients were prospectively entered into NORVASC, only patients from the denominator population were included. GPA or MPA were classified using the EMA algorithm. Survival was calculated to 31 December 2009. Ninety-one patients (53 female) and 58 MPA patients (26 female) were included. Median age at diagnosis was GPA 60.1 y, MPA 69.2 y. Cumulative follow up was GPA 978 patient y and MPA 381 patient y. Median follow up GPA 7.4 y, MPA 5.0 y. Overall mean survival was 12.3 y (GPA 13.5 y; MPA 9.9 y; P = 0.011). Survival was better for GPA than MPA (P = 0.011) (table I). There was no difference in overall survival between genders but GPA males had a better survival than MPA males (P = 0.005). There was no difference in survival between the two decades (P = 0.471), GPA (P = 0.846) MPA (P = 0.179) but a trend for GPA to have better survival and than MPA. Increasing age at diagnosis was also associated with a worse prognosis.

<table>
<thead>
<tr>
<th>Year (%)</th>
<th>GPA (%)</th>
<th>MPA (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 year</td>
<td>5 years</td>
<td>10 years</td>
</tr>
<tr>
<td>Whole cohort</td>
<td>89</td>
<td>74</td>
</tr>
<tr>
<td>CPA</td>
<td>91</td>
<td>80</td>
</tr>
<tr>
<td>MPA</td>
<td>88</td>
<td>62</td>
</tr>
<tr>
<td>1988 GPA+MPA</td>
<td>87</td>
<td>69</td>
</tr>
<tr>
<td>2000 GPA+MPA</td>
<td>93</td>
<td>78</td>
</tr>
<tr>
<td>1988 GPA</td>
<td>89</td>
<td>76</td>
</tr>
<tr>
<td>2000 GPA</td>
<td>92</td>
<td>84</td>
</tr>
<tr>
<td>1988 MPA</td>
<td>81</td>
<td>55</td>
</tr>
<tr>
<td>2000 MPA</td>
<td>93</td>
<td>68</td>
</tr>
</tbody>
</table>

Discussion—Overall survival is comparable to the EUVAS patients (88% 1 y and 78% 5 y survival). Current treatments appear to have had a modest effect on the mortality of MPA but not GPA.

Conclusion—AAV continues to have a significant long-term mortality in an unselected population.

Further reading

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