A risk score for predicting short-term incidence of death or relapse in anti-neutrophil cytoplasmic antibody-associated vasculitis

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**Introduction.** Anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV), combining granulomatosis with polyangiitis (GPA) and microscopic polyangiitis (MPA), is associated with a substantial short-term risk of relapse or death, but the factors determining short-term outcome are not well known. We aimed to develop a risk score to predict the 1-year risk of death or relapse in newly diagnosed AAV.

**Methods.** We studied patients with incident AAV enrolled in 4 international, randomized, multicenter clinical trial. A set of 22 candidate variables, all assessed at diagnosis, were considered for predicting 1-year survival or relapse risk. A stepwise approach using univariate and multivariate logistic regression models in 2000 bootstrap samples were used to identify predictors of death or relapse at 1 year. The regression coefficients computed in the final model were used to derive weights for the identified predictors. The discriminative ability of the score was evaluated by analysis of the area under the receiver operating characteristic curve (AUC).

**Results.** Among the 535 subjects, we had complete data for all 22 analyzed variables for 441 subjects. At 1-year follow-up, we recorded 44 deaths and 12 relapses. We retained 9 variables for the final risk score model: age > 60 yr (weight of 1); female sex (1); ear, nose and throat involvement (1); creatinine > 300 μmol/L (3); PR3-ANCA (1) MPO-ANCA (3); peripheral neutrophil count > 7000/mm³ (2); hemoglobin level < 10 g/dl (1) and C-reactive protein level > 10 mg/L (2). Each patient was assigned a risk score between 0 and 15. Accordingly, the 1-year risk of death or relapse was low (<5%) for 32% of patients, medium (5–20%) for 50%, and high (>20%) for 18%. The AUC for the prediction model was 0.79 (95% confidence interval 0.73–0.85).

**Conclusion.** This integrative AAV-risk prediction score may be useful in predicting a patient’s risk of death or relapse on short-term follow-up and may contribute to better risk-stratified characterization and management of AAV.

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Eosinophilic granulomatosis with polyangiitis (Churg–Strauss) (EGPA), polyarteritis nodosa (PAN) and microscopic polyangiitis (MPA) with no initial Five-Factor Score-defined poor-prognosis factors (FFS = 0): Baseline factors associated with cytotoxic agent and immunomodulator prescription

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**Introduction.** Patients with FFS = 0 EGPA, PAN or MPA [1] included in two prospective CHUSPAN trials, initially received corticosteroids (CS) alone [2,3]. Because some patients need add-on therapies (AT), baseline characteristics associated with their use were sought.

**Patients.** Patients’ data were updated in 2012. Chapel Hill definitions classified EGPA, PAN and MPA [4]. Analyzed AT were all cytotoxic agents (CA), biologics (except omalizumab), IVlg (>2 g/kg) and plasma-exchange (PE). Univariate and multivariate analyses were performed.

**Results.** The study included 193 patients (75 EGPA, 61 MPA and 57 PAN). Mean ± SD overall follow-up was 97.6 ± 39.6 months, with no difference among entities. All initially received CS alone; among 86/193 (42 PAN, 32 MPA and 30 EGPA) requiring AT (mean follow-up since CS onset: 23.3 ± 34.1 months) because CS failed (37%), relapse (52%) or CS dependency (10%). 49 received IV cyclophosphamide (CYC), 13 oral CYC, 56 azathioprine, 15 methotrexate, nine mycophenolate mofetil, seven IVlg, six PE, one infliximab and one cyclosporine. Multivariate analysis retained only baseline mononeuritis multiplex (MM) (HR = 1.81 [1.12–2.93], P = 0.02) as independently associated with AT use. AT prescription rates were comparable for the three entities. At last visit, 165/193 (85%) were alive, with 94 (57%) and 28 (17%), respectively, still on CS and/or CA or biotherapy. Overall survival (OS) was comparable for patients taking only CS vs. the others. However, patients given ≥ 1 vs. 0 AT had significantly higher Vasculitis Damage Indexes: 2.93 ± 2.09 vs. 1.96 ± 1.40 (P < 0.001), reflecting more frequent osteoporosis (33 vs. 18%, P = 0.013) or peripheral neuropathy (60 vs. 38%, P = 0.004).

**Conclusion.** Despite the good OS of baseline-FFS = 0 EGPA, PAN or MPA patients, 45% required AT, mostly those with MM whose OS was similar to that of the other FFS0 patients. This MM subpopulation might be more likely to fail on CS alone, thereby supporting prospective evaluation of their initial CA use.

**References**


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