Supplementary Table 1. Predictors at diagnosis for all-cause mortality during follow-up in non-elderly AAV patients (n = 124)

| Variable                                                                 | Univariable | Multivariable |
|--------------------------------------------------------------------------|-------------|---------------|
|                                                                          | HR          | 95% CI        | p value | HR          | 95% CI        | p value |
| **Demographic data at diagnosis**                                        |             |               |         |             |               |         |
| Age                                                                      | 1.052       | 0.987–1.121   | 0.121   |             |               |         |
| Female sex                                                               | 0.508       | 0.146–1.771   | 0.288   |             |               |         |
| **Variants (MPA vs. GPA or EGPA) at diagnosis**                          | 0.473       | 0.121–1.854   | 0.283   |             |               |         |
| **ANCA positivity at diagnosis**                                         |             |               |         |             |               |         |
| MPO-ANCA (or P-ANCA)                                                     | 0.748       | 0.188–2.974   | 0.680   |             |               |         |
| PR3-ANCA (or C-ANCA)                                                     | 2.766       | 0.783–9.767   | 0.114   |             |               |         |
| **AAV-specific indices at diagnosis**                                    |             |               |         |             |               |         |
| BVAS, ≥ 18                                                               | 1.661       | 0.219–5.147   | 0.283   |             |               |         |
| FFS, ≥ 2                                                                 | 5.994       | 1.526–23.543  | 0.010   | 3.596       | 0.429–30.100  | 0.238   |
| **Clinical manifestations at diagnosis**                                 |             |               |         |             |               |         |
| General                                                                  | 2.263       | 0.636–8.058   | 0.208   |             |               |         |
| Cutaneous                                                                | 0.122       | 0.015–1.022   | 0.052   |             |               |         |
| Muco-membranous/ocular                                                   | 1.692       | 0.213–5.592   | 0.916   |             |               |         |
| Ear nose throat                                                           | 1.973       | 0.546–7.120   | 0.300   |             |               |         |
| Pulmonary                                                                | 3.608       | 0.897–13.709  | 0.071   |             |               |         |
| Cardiovascular                                                           | 5.144       | 1.314–20.137  | 0.019   | 1.934       | 0.230–16.274  | 0.544   |
| Gastrointestinal                                                         | 1.684       | 0.134–8.735   | 0.480   |             |               |         |
| Renal                                                                    | 9.993       | 1.221–81.807  | 0.032   |             |               |         |
| Nervous                                                                  | 0.363       | 0.076–1.727   | 0.203   |             |               |         |
| **Laboratory results at diagnosis**                                      |             |               |         |             |               |         |
| White blood cell count                                                   | 1.000       | 1.000–1.000   | 0.458   |             |               |         |
| Hemoglobin                                                               | 0.945       | 0.713–1.251   | 0.691   |             |               |         |
| Platelet count                                                           | 0.999       | 0.994–1.004   | 0.689   |             |               |         |
| Fasting glucose                                                          | 1.002       | 0.984–1.021   | 0.795   |             |               |         |
| BUN                                                                      | 1.000       | 0.981–1.019   | 0.999   |             |               |         |
| Creatinine                                                               | 0.946       | 0.735–1.218   | 0.667   |             |               |         |
| Total serum protein                                                      | 0.755       | 0.380–1.499   | 0.422   |             |               |         |
| Serum albumin                                                            | 0.651       | 0.271–1.568   | 0.339   |             |               |         |
| ALP                                                                      | 1.000       | 0.992–1.009   | 0.937   |             |               |         |
| AST                                                                      | 0.924       | 0.819–1.043   | 0.202   |             |               |         |
| ALT                                                                      | 0.999       | 0.974–1.024   | 0.914   |             |               |         |
| Total bilirubin                                                          | 1.139       | 0.081–16.043  | 0.923   |             |               |         |
| ESR                                                                      | 0.995       | 0.978–1.013   | 0.592   |             |               |         |
| CRP                                                                      | 1.007       | 0.995–1.020   | 0.251   |             |               |         |

AAV, antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis; HR, hazard ratio; CI, confidence interval; MPA, microscopic polyangiitis; GPA, granulomatosis with polyangiitis; EGPA, eosinophilic granulomatosis with polyangiitis; MPO, myeloperoxidase; P, perinuclear; PR3, proteinase 3; C, cytoplasmic; BVAS, Birmingham Vasculitis Activity Score; FFS, five factor score; BUN, blood urea nitrogen; ALP, alkaline phosphatase; AST, aspartate transaminase; ALT, alanine transaminase; ESR, erythrocyte sedimentation rate; CRP, C-reactive protein.