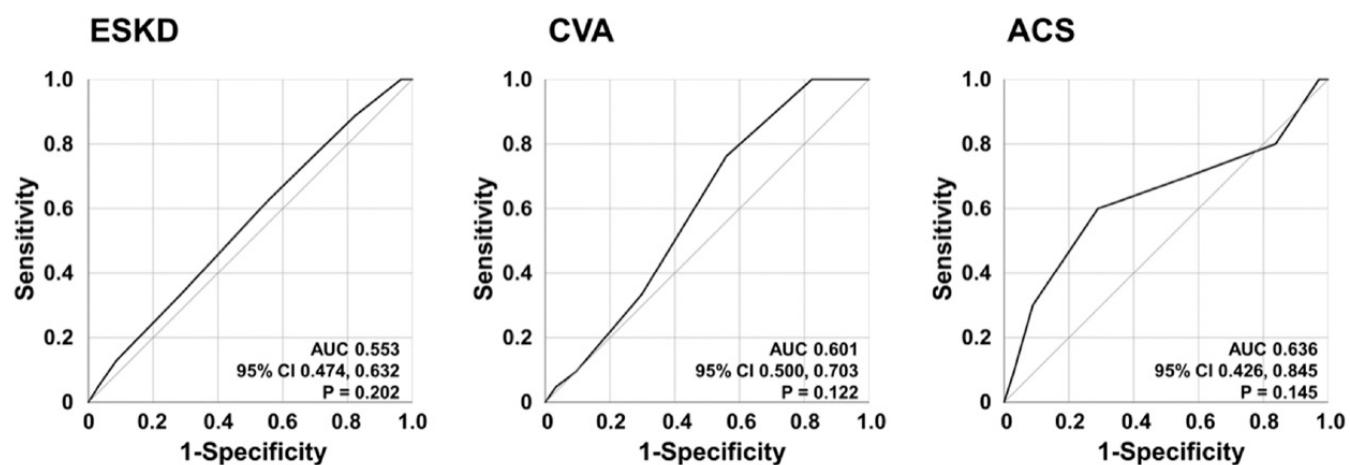
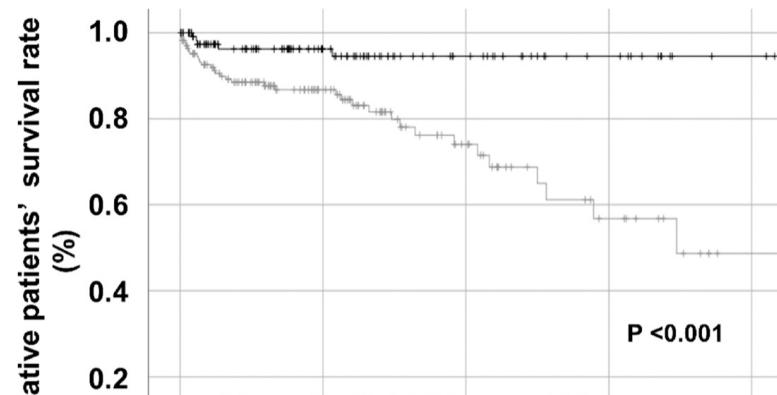


Supplementary Fig. S1. Comparison of AUCs of variables with significance in the multivariable Cox analysis for all-cause mortality.
AUC: area under the curve.

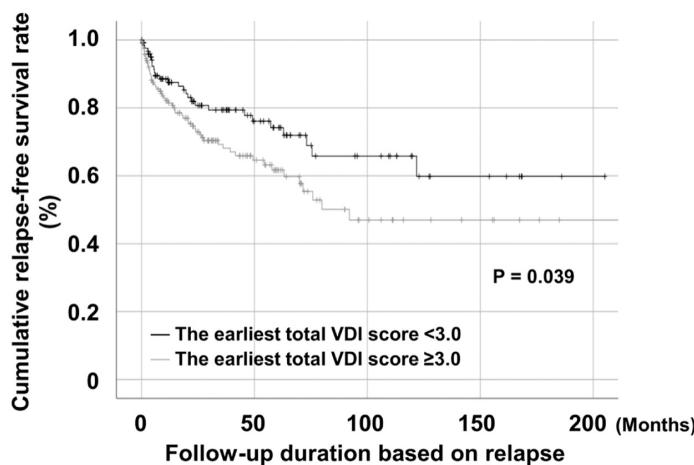


Supplementary Fig. S2. The ROC curve analysis for evaluating the predictive ability of the cut-off of the earliest total VDI score for ESKD, CVA, and ACS.

AUC: area under the curve; ROC: receiver operating characteristic; VDI: vasculitis damage index; ESKD: end-stage kidney disease; CVA: cerebrovascular accident; ACS: acute coronary syndrome.



Supplementary Fig. S3. Optimal cut-off of the earliest total VDI score for relapse and relative risk.
AUC: area under the curve; RR: relative risk; VDI: vasculitis damage index.

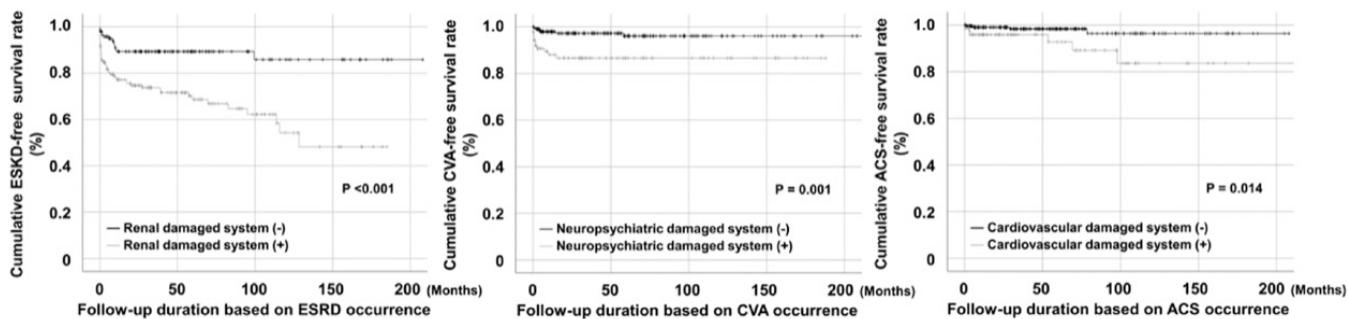


Supplementary Fig. S4. Comparison of cumulative relapse-free survival rates according to the earliest total VDI scores ≥ 3.0 .

Supplementary Table S1. Cox hazards model analyses of the earliest VDI with variables at diagnosis for relapse during follow-up in AAV patients.

Variables	Univariable			Multivariable (VDI)			Multivariable (VID ≥ 3.0)		
	HR	95% CI	p value	HR	95% CI	p value	HR	95% CI	p value
Age (years)	0.985	0.971, 0.999	0.031	0.982	0.967, 0.997	0.017	0.981	0.966, 0.996	0.013
Male gender (N, (%))	1.233	0.787, 1.931	0.360						
BMI (kg/m^2)	0.932	0.869, 1.000	0.050						
Ex-smoker (N, (%))	1.197	0.378, 3.794	0.760						
MPO-ANCA (or P-ANCA) positivity	1.088	0.689, 1.716	0.718						
PR3-ANCA (or C-ANCA) positivity	2.516	1.573, 4.027	<0.001	2.220	1.337, 3.685	0.002	2.255	1.364, 3.730	0.002
BVAS	1.030	0.998, 1.063	0.067						
FFS	1.105	0.898, 1.361	0.345						
DM	0.766	0.463, 1.267	0.299						
Hypertension	1.547	1.008, 2.374	0.046	1.534	0.993, 2.371	0.054	1.510	0.976, 2.338	0.064
Dyslipidaemia	1.131	0.678, 1.888	0.637						
White blood cell count ($/\text{mm}^3$)	1.000	1.000, 1.000	0.929						
Haemoglobin (g/dL)	0.896	0.815, 0.985	0.024	0.918	0.823, 1.023	0.122	0.914	0.821, 1.018	0.101
Platelet count ($\times 1000/\text{mm}^3$)	0.999	0.998, 1.001	0.461						
Fasting glucose (mg/dL)	1.002	0.998, 1.007	0.328						
Blood urea nitrogen (mg/dL)	1.013	1.003, 1.024	0.009	1.011	1.000, 1.022	0.047	1.011	1.001, 1.022	0.038
Serum creatinine (mg/dL)	1.069	0.965, 1.183	0.202						
Protein (g/dL)	0.919	0.747, 1.132	0.428						
Serum albumin (g/dL)	0.842	0.640, 1.109	0.221						
ESR (mm/hr)	1.003	0.998, 1.009	0.191						
CRP (mg/L)	1.002	0.998, 1.005	0.317						
VDI	1.202	1.030, 1.402	0.020	1.122	0.959, 1.311	0.150			
VID ≥ 3.0	1.605	1.019, 2.529	0.041				1.440	0.909, 2.283	0.120

VDI: vascular damage index; AAV: ANCA-associated vasculitis; ANCA: antineutrophil cytoplasmic antibody; BMI: body mass index; MPO: myeloperoxidase; P: perinuclear; PR3: proteinase 3; C: cytoplasmic; BVAS: Birmingham vasculitis activity score; FFS: five-factor score; DM: diabetes mellitus; DM: diabetes mellitus; ESR: erythrocyte sedimentation rate; CRP: C-reactive protein.



Supplementary Fig. S5. Association of the presence of each damaged system comprising the earliest total VDI score with each damaged system-related complication.

ESKD: end-stage kidney disease; CVA: cerebrovascular accident; ACS: acute coronary syndrome; VDI: vasculitis damage index.