

**Supplementary Table I.** Comparison of variables between survived and deceased patients with AAV.

Variables	Survived patients (n=139)	Deceased patients (n=14)	p-value
<i>Demographic data</i>			
Age at diagnosis (year old)	54.9 ± 15.1	57.9 ± 16.7	0.487
Male gender (n, (%))	41 (29.5)	6 (42.9)	0.302
Follow-up duration (months)	51.0 ± 48.5	56.9 ± 59.5	0.673
<i>Variants of AAV at diagnosis (n, (%))*</i>			
MPA	76 (54.7)	7 (50.0)	
GPA	30 (21.6)	7 (50.0)	
EGPA	33 (23.7)	0 (0)	
MPA vs. GPA			0.098
MPA vs. EGPA			0.085
GPA vs. EGPA			0.008
<i>Clinical manifestations at diagnosis (n, (%))</i>			
General manifestation	61 (43.9)	7 (50.0)	0.661
Cutaneous manifestation	34 (24.5)	2 (14.3)	0.392
Mucous membranes/Eyes manifestation	10 (7.2)	2 (14.3)	0.347
Ear Nose Throat manifestation	49 (35.3)	5 (35.7)	0.972
Cardiovascular manifestation	33 (23.7)	12 (85.7)	<0.001
Gastrointestinal manifestation	8 (5.8)	2 (14.3)	0.218
Pulmonary manifestation	69 (49.6)	11 (78.6)	0.039
Renal manifestation	79 (56.8)	11 (78.6)	0.115
Nervous systemic manifestation	48 (34.5)	4 (28.6)	0.654
<i>ANCA at diagnosis (n, (%))</i>			
MPO-ANCA (or P-ANCA)	86 (61.9)	8 (57.1)	0.729
PR3-ANCA (or C-ANCA)	22 (15.8)	4 (28.6)	0.226
Both ANCAs	6 (4.3)	1 (7.1)	0.630
ANCA negative	37 (26.6)	3 (21.4)	0.674
BVAS or BVAS for GPA at diagnosis	12.1 ± 7.6	13.2 ± 7.7	0.592
FFS (2009) at diagnosis	1.1 ± 0.9	2.5 ± 0.8	<0.001
<i>Comorbidities at diagnosis (n, (%))</i>			
CKD stage ≥ 3	52 (37.4)	12 (85.7)	<0.001
DAH	8 (5.8)	4 (28.6)	0.002
<i>Comorbidities during the follow-up (n, (%))</i>			
ESRD	20 (14.4)	4 (28.6)	0.164
DM	28 (20.1)	6 (42.9)	0.051
HTN	56 (40.3)	11 (78.6)	0.006
ILD	53 (38.1)	10 (71.4)	0.016
Hyperthyroidism	8 (5.8)	3 (21.4)	0.030
Hypothyroidism	4 (2.9)	0 (0)	0.520
IHD	8 (5.8)	2 (14.3)	0.218
CVA or carotid arterial stenosis	12 (8.6)	3 (21.4)	0.125
HBV or HCV carrier	4 (2.9)	1 (7.1)	0.392
Other rheumatic diseases	15 (10.8)	0 (0)	0.196
<i>Medications or procedure during the follow-up (n, (%))</i>			
GC	119 (85.6)	13 (92.9)	0.453
CYC	56 (40.3)	9 (64.3)	0.083
MMF	8 (5.8)	2 (14.3)	0.218
AZA	41 (29.5)	1 (7.1)	0.074
CNI	9 (6.5)	0 (0)	0.326
RTX	12 (8.6)	3 (21.4)	0.125
MTX	13 (9.4)	0 (0)	0.232
PE	3 (2.2)	1 (7.1)	0.265
Relapse during the follow-up (n, (%))	36 (25.9)	8 (57.1)	0.014

Values are expressed as mean and standard deviation or N (%).

AAV: antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis; 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; CKD: chronic kidney disease; DAH: diffuse alveolar haemorrhage; ESRD: end stage renal disease; DM: diabetes mellitus; HTN: hypertension; ILD: interstitial lung disease; IHD: ischaemic heart disease; CVA: cerebrovascular accident; GC: glucocorticoid; CYC: cyclophosphamide; MMF: mycophenolate mofetil; AZA: azathioprine; CNI: calcineurin inhibitor; RTX: Rituximab; MTX: methotrexate; PE: plasma exchange.