Supplementary Table 1. Percentages of tissue and organ involvement at the time of diagnosis in patients with AAV: data from the DCVAS and TRVaS cohorts

	DCVAS	TRVaS	DCVAS	TRVaS	DCVAS	TRVaS
System involvement, %	**GPA (n=735)**	**GPA (n=785)**	**MPA (n=334)**	**MPA (n=191)**	**EGPA (n=255)**	**EGPA (n=164)**
Constitutional symptoms	77.7	85	86.5	77	79.6	69
Cutaneous involvement	32.2	37	26	34	49.4	58.5
Ocular involvement	38.6	38	13.7	29	15.3	38.4
ENT involvement	80.4	71	25.7	31	72.5	56.1
Pulmonary involvement	66.8	80	67.4	75	89.4	76.8
Cardiac involvement	11.1	25	15.5	28	49.4	43.3
Renal involvement	69.8	69	90.1	92	37.6	42.1
Gastrointestinal involvement	19	26	22.1	26	30.6	36.4
Neurological involvement	20.3	33	26.3	33	56	57.3

Supplementary Table 2. Various classification and scoring systems of renal biopsies in AAV

Berden classification

Classification	Definition
Focal	≥50% normal glomeruli in renal biopsy
Crescentic	≥50% of glomeruli showing cellular crescents in renal biopsy
Mixed	Presence of a combination of both focal and crescentic formations
Sclerotic	≥50% of glomeruli showing sclerosis in renal biopsy

Mayo Clinic/Renal Pathology Society Chronicity Score

Glomerular sclerosis	Tubular atrophy	Interstitial fibrosis	Arteriosclerosis
<%10=0	<%10=0	<%10=0	-
%10-25=1	%10-25=1	%10-25=1	Intimal thickness ≥ medial thickness = 1 point
%26-50=2	%26-50=2	%26-50=2	-
>%50=3 point	>%50=3 point	>%50=3 point	-

Score: ranges from 0 to 10 and is divided into four risk groups
Minimal (0-1), mild (2-4), moderate (5-7), and severe (≥8)

Renal risk scoring system

Normal glomeruli	Tubular atrophy and interstitial fibrosis	Baseline estimated glomerular filtration rate (eGFR)
>%25 = 0	≤%25 = 0	>15 ml/dk/1.73 m² = 0
%10-25= 4	>%25=2 point	≤15 ml/dk/1.73 m²= 3 point
<%10= 6 point		

Score: ranges from 0 to 11 and is divided into three risk groups
Low (0-1 points), medium (2-7 points), and high (8-11 points) risk of ESRD

Supplementary Table 3. European Alliance of Associations for Rheumatology (EULAR) consensus terms for treatment response in patients with AAV

EULAR definitions of disease states
Active disease: Presence of symptoms and signs attributable to active AAV (e.g., glomerulonephritis, pulmonary nodules).
Remission: Absence of typical symptoms and signs of AAV (may or may not be under immunosuppressive therapy).
Sustained remission: Absence of typical symptoms and signs of AAV for a defined period of time (may or may not be under immunosuppressive therapy).
Treatment response: ≥50% reduction in disease activity score without the appearance of new manifestations.
Relapse: Recurrence of active AAV following a period of remission.
Refractory disease: Persistence or worsening of active AAV symptoms and signs despite continuation of standard induction therapy for a defined period (damage, infection, and treatment-related comorbidities should be excluded).