# Flowchart A | Study design with recruitment sites and patient separation ANCA patients (N = 1591) Angers (n=173) Baltimore (n=148) Irish Registry (n=224) London (n=171)Manchester (n=128) Mexico City (n=89) Prague (n=62) Preston (n=72) Salford (n=100) Scottish Registry (n=360) Uludağ (n=65) Excluded (N = 152)Insufficient follow-up (n=77) Missing data (n = 75) Analyzed (N = 1439) Validation (N=480) Development (N=959) Flowchart B | patient flow through study Analyzed (N=1439) n=291 RRT at time of diagnosis(n=291) n=136 n=141 n=155 Renal recovery (n=136) n=29 n=831 ESKD (n=325)

n=84

Alive at last F/U (n=1124)

n=209

n=23

n=176

Died (n=315)

n=116

# Supplementary Table 1 | Basic characteristics and outcome of complete cohort, development, and validation cohorts

	Development Cohort	Validation Cohort	=
	(N = 959)	(N = 480)	Р
Age at diagnosis (yr)	63.4 (52.3-72.5)	64.5 (53.9-73.2)	0.449
Sex Male	484 (50.5%)	266 (55.4%)	0.086
Diagnosis			
GPA	388 (40.5%)	180 (37.5%)	0.588
MPA	538 (56.1%)	282 (58.8%)	
EGPA	14 (1.5%)	5 (1.0%)	
ANCA Negative	19 (2.0%)	12 (2.5%)	
ANCA type			
Myeloperoxidase	509 (53.1%)	253 (52.7%)	0.626
Proteinase 3	389 (40.6%)	190 (39.6%)	
ANCA Negative	61 (6.4%)	37 (7.7%)	
eGFR (ml/min/1.73m²)	21.8 (11.3-38.0)	18.0 (9.8-35.0)	0.026
Creatinine (µmol/L)	225.0 (147.0-383.5)	257.0 (157.5-449.5)	0.015
Initial Need for RRT	185 (19.3%)	106 (22.1%)	0.240
Glomeruli on biopsy	17.0 (12.0-23.0)	17.0 (12.0-23.2)	0.718
Percentage Normal Glomeruli	36.0 (14.3-66.7)	34.7 (14.3-62.5)	0.296
Percentage Crescentic Glomeruli	19.0 (5.8-44.4)	21.4 (6.3-50.0)	0.292
IFTA			
None - Mild	469 (48.9%)	220 (45.8%)	0.297
Mild to Moderate - Severe	490 (51.1%)	260 (54.2%)	
Renal Risk Score	3.0 (0.0-7.0)	4.0 (2.0-7.0)	0.150
Risk Group			
Low	258 (26.9%)	112 (23.3%)	
Moderate	484 (50.5%)	258 (53.8%)	
High	217 (22.6%)	110 (22.9%)	

Data presented as n (%) or median (interquartile range). The eGFR was calculated using the Chronic Kidney Disease Epidemiology Collaboration (CKD EPI) equation. ANCA, anti-neutrophil cytoplasmic antibody; eGFR, estimated glomerular filtration rate; EGPA, eosinophilic granulomatosis with polyangiitis; GPA, granulomatosis with polyangiitis; IFTA, interstitial fibrosis and tubular atrophy; MPA, microscopic polyangiitis; RRT, renal replacement therapy.

## **Supplementary Table 2 | Clinical outcomes**

	Development	Validation	
	Cohort	Cohort	
	(N=959)	(N=480)	Р
Follow-up (years)	3.6 (1.1-5.9)	3.6 (1.0-6.0)	0.770
Renal recovery	83/185 (44.9%)	53/106 (50%)	0.681
ESKD	207 (14.4%)	118 (8.2%)	0.254
Mortality	206 (14.3%)	109 (7.6%)	0.504

# Supplementary Table 3 | Validated 3-year survival

	Original 3-year	Validated 3-year
Group	renal survival	renal Survival
Low	100%	94.8%
Medium	84%	86.1%
High	32%	52.4%

# Supplementary Table 4 | Parameters of the continuous model

		β
Creatinine	Log(µmol)	1.250
Normal Glomeruli	%	-0.0167
IFTA	≥25%	0.616
	C Statistic	0.833

Survival function in supplementary Equation 1; model valid up to t=10 years; log, natural logarithm

Supplementary Table 5 | Patient cohorts divided by endpoint

	No ESKD	ESKD
	(N = 752)	(N = 207)
Age at diagnosis (yr)	64.1 (53.9-73.1)	62.0 (50.5-70.9)
Sex Male	567 (50.9%)	183 (56.5%)
Diagnosis		
GPA	443 (39.8%)	125 (38.5%)
MPA	632 (56.7%)	188 (57.8%)
EGPA	16 (1.4%)	3 (0.9%)
ANCA Negative	22 (2.0%)	9 (2.8%)
ANCA type		
Myeloperoxidase	509 (53.1%)	253 (52.7%)
Proteinase 3	389 (40.6%)	190 (39.6%)
ANCA Negative	61 (6.4%)	37 (7.7%)
eGFR (ml/min/1.73m²)	25.0 (14.0-41.3)	9.0 (5.8-16.0)
Creatinine (µmol/L)	201.6 (136.0-314.7)	471.0 (300.6-708.0)
Initial Need for RRT	108 (9.7%)	183 (56.5%)
Glomeruli on biopsy	17.0 (12.0-24.0)	16.0 (12.0-22.0)
Percentage Normal Glomeruli	42.9 (22.2-70.0)	13.3 (0.0-33.3)
Percentage Crescentic Glomeruli	20.0 (5.6-43.4)	24.0 (7.1-51.9)
TA/IF		
None - Mild	587 (52.6%)	102 (31.5%)
Mild to Moderate - Severe	528 (47.4%)	222 (68.5%)
Renal Risk Score	2.0 (0.0-5.0)	8.0 (5.0-11.0)
Risk Group		
Low	348 (31.2%)	22 (6.8%)
Moderate	610 (54.8%)	132 (40.6%)
High	156 (14.0%)	171 (52.6%)

Data presented as n (%) or median (interquartile range). The eGFR was calculated using the Chronic Kidney Disease Epidemiology Collaboration (CKD EPI) equation. ANCA, antineutrophil cytoplasmic antibody; eGFR, estimated glomerular filtration rate; EGPA, eosinophilic granulomatosis with polyangiitis; GPA, granulomatosis with polyangiitis; IFTA, interstitial fibrosis and tubular atrophy; MPA, microscopic polyangiitis; RRT, renal replacement therapy.

## **Supplementary Table 6 | Univariable models for ESKD**

	- a		
	HR	95% CI	Р
Age	0.998	(0.990-1.005)	0.500
Sex: Male	0.846	(0.680-1.054)	0.135
Antibody: PR3	0.940	(0.744-1.187)	0.601
eGFR	0.937	(0.927-0.947)	<0.001
Log(Creatinine)	4.524	(3.847-5.320)	<0.001
Percentage Normal Glomeruli	0.970	(0.965-0.975)	<0.001
Percentage Crescentic Glomeruli	1.006	(1.002-1.010)	0.005
IFTA: mild to Moderate - Severe	2.307	(1.824-2.917)	<0.001

Reference categories: Female, GPA, MPO, None-Mild. CI, confidence interval; eGFR, estimated glomerular filtration rate; GPA, granulomatosis with polyangiitis; HR, hazard ratio; IFTA, interstitial fibrosis and tubular atrophy; log, natural logarithm; MPA, microscopic polyangiitis; MPO, myeloperoxidase; PR3, proteinase 3.

#### **Supplementary Table 7 | Multivariable for ESKD**

	HR	CI	Р
Age	1.010	(0.998-1.022)	0.108
Sex: Male	1.361	(0.960-1.929)	0.083
Antibody: PR3	0.847	(0.602-1.191)	0.340

	HR	CI	P
eGFR	1.029	(1.013-1.045)	<0.001
Log(Creatinine)	5.918	(3.909-8.960)	< 0.001
Percentage Normal Glomeruli	0.974	(0.964-0.984)	< 0.001
Percentage Crescentic Glomeruli	0.996	(0.991-1.002)	0.197
TA/IF: Mild to Moderate - Severe	1.735	(1.196-2.518)	0.004

Reference categories: Female, GPA, MPO, None-Mild. CI, confidence interval; eGFR, estimated glomerular filtration rate; GPA, granulomatosis with polyangiitis; HR, hazard ratio; IFTA, interstitial fibrosis and tubular atrophy; log, natural logarithm; MPA, microscopic polyangiitis; MPO, myeloperoxidase; PR3, proteinase 3.