

Supplemental table 1. Demographics and baselines characteristics of patients with normal and low sC3 at diagnosis.

Demographics	Normal sC3	Low sC3	p-value
Disease related Parameters	N=91	N=24	
N (%) or mean (SD)			
Vasculitic manifestations			
General Symptoms	55 (60.5)	11 (45.8)	0.19
Fever	48 (53.3)	11 (45.8)	0.51
Arthralgias	48 (52.3)	9 (37.5)	0.18
Myalgias	37 (40.7)	10 (41.7)	0.92
Weight Loss	6 (6.6)	3 (12.5)	0.33
Flu Like Syndrome	48 (52.7)	8 (33.3)	0.09
Cutaneous Involvement	12 (13.2)	2 (8.3)	0.73
Mucus Involvement	4 (4.4)	0 (0)	0.57
Eye Involvement	2 (2.2)	0 (0)	1
Ear nose throat Involvement	26 (28.6)	4 (16.7)	0.30
Lung Involvement	36 (39.6)	12 (50)	0.35
Gastrointestinal Involvement	3 (3.3)	0 (0)	1
Neurological Involvement	3 (3.3)	1 (4.2)	1
estimated GFR (ml/min/1.73m ²)			0.5
<30 ml/min/1.73m ²	71(7.8)	20 (83.3)	
>30 ml/min/1.73m ²	20 (2.2)	4 (16.7)	
Number of Glomeruli (median)	20(12)	16(8)	0.11

Supplemental Table 2. Univariate analysis for Risk factors associated with the composite outcome of ESKD or death at the end of the 1 st year after PIGN diagnosis.		
Variable	ESKD or death	
Univariate model	OR (95% CI)	p value
Age	1.08 (0.99-1.05)	0.08
Gender female	0.8 (0.32-2.01)	0.64
Caucasian race vs. all others	0.23 (0.03-1.75)	0.15
Low serum C3 (vs normal serum C3)	4.28 (1.57-1.67)	0.004
PR3-ANCA vs. MPO-ANCA	0.41 (0.14-1.22)	0.11
Microscopic Polyangiitis vs. Renal Limited Disease	1.11 (0.41-2.99)	0.82
Granulomatosis with polyangiitis vs. Renal Limited Disease	0.13 (0.05-1.45)	0.28
Serum creatinine (mg/dL)	1.38 (1.15-1.66)	<0.001
estimated GFR (ml/min/1.73m ²)	0.88 (0.82-0.96)	0.003
estimated GFR > 30 vs. <30 ml/min/1.73m ²	0.13 (0.017-1.06)	0.058
Oliguria	10.85 (3.38-34.84)	<0.01
Acute dialysis requirement	7.7 (2.83-20.9)	<0.001
Vasculitic manifestation		
General symptoms	1.19 (0.47-3.04)	0.70
Fever	1.44 (0.56-3.70)	0.44
Arthralgias	1.41 (0.56-3.55)	0.45
Myalgias	1.77 (0.70-4.45)	0.22
Weight loss	0.47 (0.05 – 4.02)	0.49
Flue like syndrome	0.95 (0.38 – 2.38)	0.92
Cutaneous involvement	1.10 (0.28-4.33)	0.88
Mucus involvement	1.34 (0.13 – 13.5)	0.80
Ear nose throat involvement	0.53 (0.16 – 1.72)	0.29
Pulmonary involvement	0.87 (0.34 – 2.22)	0.77
BVAS score	1.0 (0.92-1.08)	0.99
Histopathological parameters		
Normal glomeruli >50% vs.< 50%	0.8 (0.67-0.95)	0.01

Normal glomeruli 25-50% vs. >50%	2.8 (0.29-26.4)	0.36
Normal glomeruli 0-25% vs. >50%	4.28 (0.51-35.3)	0.17
Normal glomeruli >10% vs. <10%	0.4 (0.15-1.06)	0.06
EUVAS categorization		
Focal class (vs. Sclerotic class)	0.05 (0.006-0.54)	0.01
Crescentic class (vs. Sclerotic class)	0.32 (0.08-1.27)	0.10
Mixed class (vs. Sclerotic class)	0.35 (0.10-1.23)	0.10
Arteriosclerosis moderate or severe vs. none/mild	4.71 (1.18-18.79)	0.02
Interstitial fibrosis, moderate or severe vs. none/mild	2.1 (0.16-1.63)	0.26
Tubular atrophy moderate or severe vs. none/mild	1.37 (0.51-3.64)	0.52
Fibrotic crescents/segmental sclerosis >25% vs. none/mild	2.03 (0.54-7.54)	0.29
Global glomerulosclerosis moderate or severe vs. none/mild	3.0 (0.96-9.39)	0.05
Chronicity score (all)	1.74 (1.30-2.34)	<0.01
Chronicity score (8-12) vs. (1-7)	4.44 (1.61-12.25)	0.004
Cellular Crescents >25-50% vs. <25%	2.99 (0.87-10.2)	0.08
Cellular Crescents >50% vs. <25%	3.27 (0.95-11.2)	0.059
Glomerular Necrosis >25-50% vs. <25%	0.17 (0.022-1.43)	0.10
Glomerular Necrosis >50% vs. <25%	1.79 (0.40-7.93)	0.44
Interstitial Leucocyte Infiltration (moderate or severe vs. none/mild)	1.48 (0.5-4.40)	0.47
Interstitial Leucocyte Infiltration (severe >50% vs. none/mild)	2.35 (0.19-27.8)	0.49
Circumferential Crescents (any) present	1.56 (0.59-4.08)	0.36
Red Blood Cell Cast present in biopsy (vs. absent)	1.5 (0.41-3.96)	0.41
Fibrinoid necrosis in the vessel wall present (vs. absent)	1.03 (0.20-5.29)	0.96
Activity score (all)	1.28 (0.99-1.64)	0.055
Activity score (8-12) vs. (1-7)	4.44 (1.61-12.25)	0.004
C3 deposition (any, vs. absence)	1.02 (0.28-3.63)	0.97
C3 deposition (1+ vs. absence)	0.95 (0.34-2.60)	0.92
C3 deposition (2+ vs. 1+)	0.94 (0.17-4.99)	0.94

Supplemental Table 3. Risk factors for ESKD in long-term among patients with PIGN who responded to initial treatment.

Variable (at diagnosis)	Hazard Ratio (95% CI)	p value
Univariate model		
Low serum C3	2.95 (0.95 – 9.17)	0.06
Serum creatinine	1.24 (1.04-1.48)	0.01
Oliguria	1.47 (0.57-3.80)	0.41
estimated GFR >30 vs. <30 ml/min/1.73m ²	0.23 (0.03-1.74)	0.15
Acute dialysis requirement	1.33 (0.49-3.58)	0.57
EUVAS category		
Focal class vs Sclerotic class	0.06 (0.01-0.39)	0.003
Crescentic class vs Sclerotic class	0.33 (0.09-1.14)	0.08
Mixed class vs Sclerotic class	0.19 (0.05-0.75)	0.01
Normal glomeruli >10% (versus <=10%)	0.39 (0.13-1.16)	0.09
Interstitial fibrosis (moderate or severe vs. none/mild)	3.08 (1.14-8.31)	0.02
RBC cast present in specimen	0.16 (0.03-0.73)	0.01
C3 1+ or more by IFF	1.9 (0.64-5.58)	0.24
Activity Index	1.09 (0.84-1.40)	0.50
Chronicity Index	1.30 (0.96-1.77)	0.08

Supplemental Table 4. Univariate analysis for determination of predictors for response to initial immunosuppressive therapy in patients with PIGN.

Variable		
Univariate model	Odds ratio (95% CI)	p-value
Age (years)	0.98 (0.95-1.01)	0.24
Gender (males)	1.27 (0.49-3.31)	0.6
Caucasian Race	4.84 (0.64-36.54)	0.12
ANCA type C/PR3-ANCA vs. P/MPO-ANCA	2.05 (0.68-6.12)	0.11
Clinical phenotype		
Granulomatosis with Polyangiitis (vs. Renal Limited Disease)	3.48 (0.68-17.64)	0.13
Microscopic Polyangiitis (vs. Renal Limited Disease)	1.13 (0.40-3.12)	0.81
Oliguria	0.07 (0.02-0.27)	<0.001
estimated GFR >30 vs. <30 ml/min/1.73m ²	6.47 (0.82-50.9)	0.07
Serum creatinine (mg/dL)	0.7 (0.58-0.85)	<0.001
Acute dialysis requirement	0.09 (0.03-0.27)	<0.01
Ear Nose Throat involvement	1.62 (0.49-5.28)	0.42
Pulmonary involvement	0.94 (0.36-2.46)	0.9
Skin involvement	3.20 (0.39-26.0)	0.72
BVAS score	0.98 (0.91-1.07)	0.80
Low serum C3	0.19 (0.06-0.53)	0.002
Normal glomeruli >10% (vs. <=10%)	2.8 (1.04-7.55)	0.04
Arteriosclerosis, (severe vs. mild/moderate)	0.26 (0.06-1.09)	0.06
Interstitial fibrosis (severe vs. mild/moderate)	0.47 (0.07-2.89)	0.42
Tubular atrophy, (severe vs .mild/moderate)	0.83 (0.08-8.26)	0.87
Fibrotic crescents/segmental sclerosis >50%	0.45 (0.12-1.68)	0.23
Global glomerulosclerosis >50% glomeruli	0.43 (0.12-1.5)	0.18
Chronicity score per unite increase	0.57 (0.42-0.76)	0.001

Cellular crescents >50%	0.36 (0.10-1.28)	0.11
Glomerular necrosis severe vs. mild/moderate	0.55 (0.12-2.46)	0.44
Interstitial Leucocyte Infiltration (severe vs. mild/moderate)	0.61 (0.20-1.83)	0.38
Circumferential Crescents (present vs. absent)	0.57 (0.21-1.52)	0.26
Activity score (all)	0.79 (0.61-1.02)	0.08
Activity score (8-12) vs. (1-7)	0.35 (0.09-1.33)	0.12
Chronicity score (all)	0.57 (0.42-0.76)	<0.001
Chronicity score 8-12 vs. 1-7	0.25 (0.09-0.69)	0.008
C3 staining (1+ vs. 2+)	0.65 (0.10-4.13)	0.65
C3 staining (any)	0.97 (0.27-3.45)	0.97
EUVAS categories		
Focal class (vs. Sclerotic class)	16.8 (1.82-154.9)	0.01
Crescentic class (vs. Sclerotic class)	4.02 (0.95-16.9)	0.057
Mixed class (vs. Sclerotic class)	2.8 (0.81-9.65)	0.1