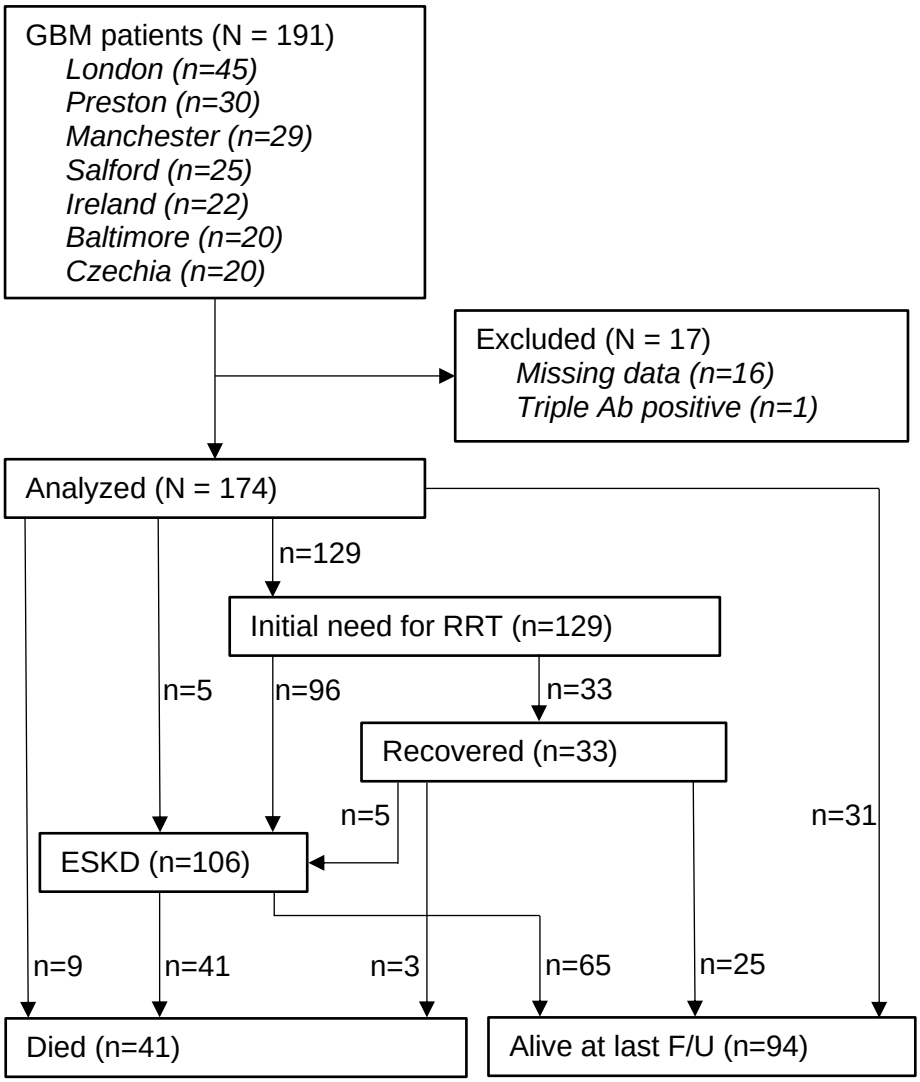
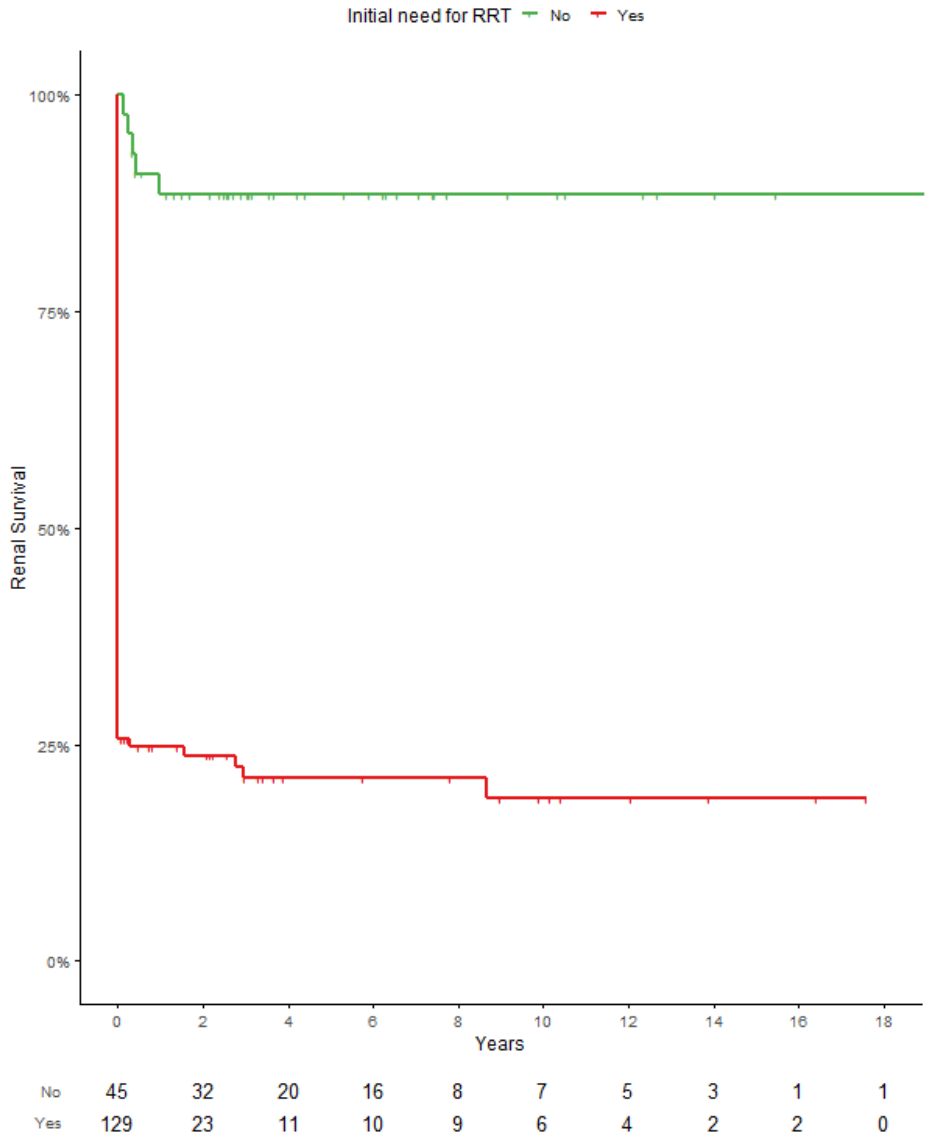
Supplemental Table of Contents:

SUPPLEMENTAL FIGURES

Supplemental Figure 1 | Flow chart diagram

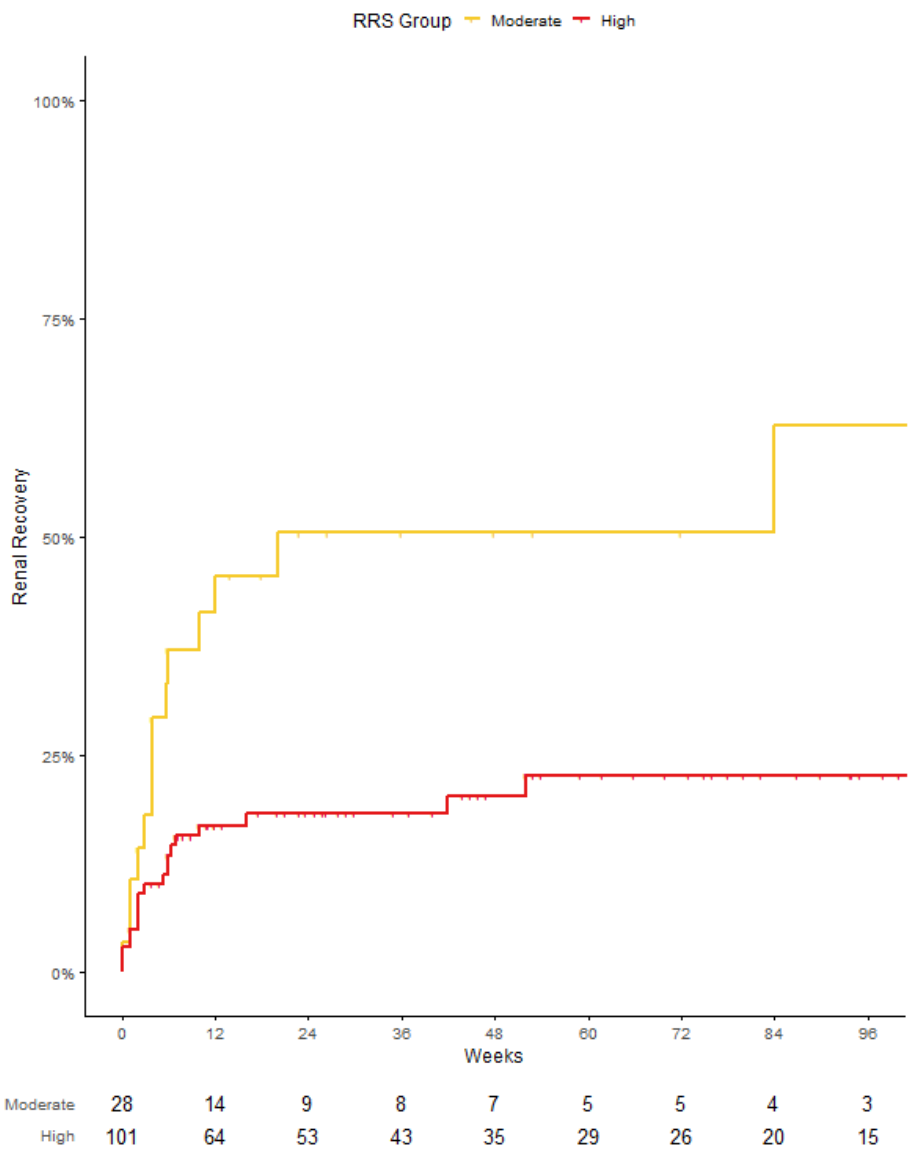


Supplemental Figure 2



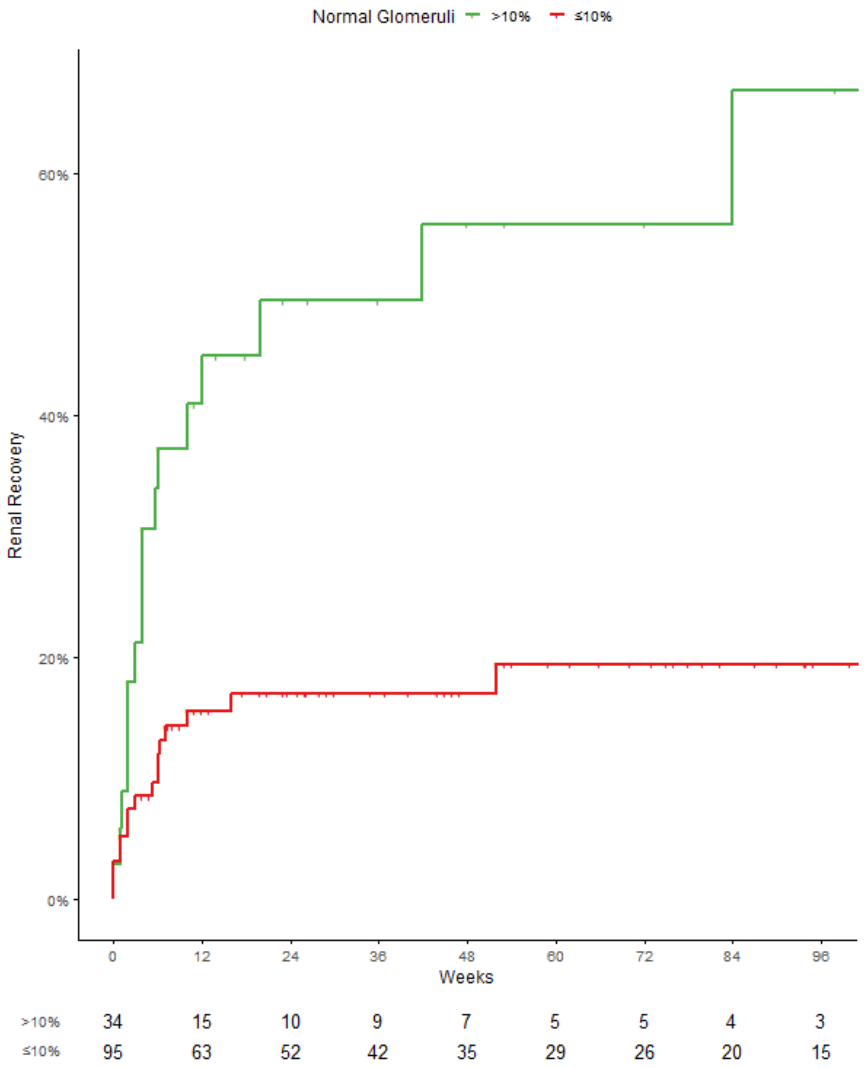
Supplemental Figure 2 | Kidney survival according to the initial need for renal replacement therapy. Kaplan-Meier curve demonstrating the development of end stage kidney disease (ESKD) of patients with anti-glomerular basement membrane (GBM) disease. ESKD differs between the two patient groups, *C*=0.767, *P*<0.001.

Supplemental Figure 3a



Supplemental Figure 3a | Recovery of kidney function according to the Renal Risk Score. Kaplan-Meier curve demonstrating renal recovery of patients with anti-glomerular basement membrane (GBM) disease and initially requiring renal replacement therapy (RRT) according to their Renal Risk Score (RRS) group (low, 0 points; moderate, 2–7 points; high, 8–11 points). Patients in the moderate risk group recovered kidney function more often compared to patients in the high risk group, *C*=0.644, *P*<0.001.

Supplemental Figure 3b



Supplemental Figure 3b | Recovery of kidney function according to the percentage of normal glomeruli in the kidney biopsy. Kaplan-Meier curve demonstrating renal recovery of patients with anti-glomerular basement membrane (GBM) disease and initially requiring renal replacement therapy grouped by the percentage normal glomeruli on their biopsy (Normal ≥ 10%, Normal < 10%). Patients with more than 10% normal glomeruli in the biopsy recovered renal function more often compared to patients with a lower percentage of normal glomeruli than 10%, Harrell's *C*=0.622, *P*<0.001.

SUPPLEMENTARY TABLES

Supplementary Table 1 | Therapeutic Intervention

Therapies	Patients (N=174)
All Therapies	131 (75.3%)
Glucocorticoids	156 (89.7%)
Cyclophosphamide	146 (83.9%)
Plasma Exchange	143 (82.2%)
No Therapies	7 (4.0%)

All therapies, patients receiving glucocorticoids, cyclophosphamide, and plasma exchange; no therapies, patients who did not receive any therapeutic intervention.

Supplementary Table 2a | Univariable models for time to ESKD

	HR (95% CI)	*C*	*P* Value
Ab (Mono Anti-GBM, Double Positive Ab)	0.870 (0.653 - 1.153)	0.538	0.333
Female Sex	0.893 (0.681 - 1.170)	0.530	0.412
Age at diagnosis	1.013 (1.002 - 1.025)	0.588	0.024
eGFR	0.897 (0.857 - 0.939)	0.772	<0.001
Normal Glomeruli	0.953 (0.936 - 0.971)	0.718	<0.001
Tubular Atrophy and Interstitial Fibrosis	1.216 (0.808 - 1.828)	0.523	0.348
Initial need for RRT	14.05 (5.700 - 34.64)	0.767	<0.001
RRS	1.326 (1.207 - 1.456)	0.760	<0.001

Ab, antibody; C, Harrell's Concordance; double positive Ab, antibodies for anti-glomerular basement membrane (GBM) and antineutrophil cytoplasmic (ANCA) antibodies; C, Harrell's Concordance; eGFR, estimated glomerular filtration rate; ESKD, end stage kidney disease; HR, Hazard ratio; 95% CI, 95% confidence interval; mono anti-GBM, anti-GBM antibodies without additional ANCA antibodies; RRS, Renal Risk Score comprised of estimated glomerular filtration rate (G0>15 ml/min/1.73 m^2, G1≤15 ml/min/1.73 m^2), percentage of normal glomeruli in the kidney biopsy (N0>25%, N1=10-25%, N2<10%), tubular atrophy and interstitial fibrosis (T0 ≤ mild to moderate, T1 ≥ moderate), with points assigned (G1=3, N1=4, N2=6, T1=2) and risk groups created (low 0, moderate 2 - 7, and high risk group 8 – 11 points); RRT, renal replacement therapy.

Supplementary Table 2b | Multivariable models for time to ESKD in sub-cohort of patients with serum creatinine at presentation (n=119)

Variable	β Coefficient	HR (95% CI)	*P* Value
Ab (Mono Anti-GBM, Double Positive Ab)	-0.281	0.755 (0.509-1.121)	0.163
Female Sex	-0.078	0.925 (0.629-1.360)	0.691
Age at diagnosis	0.006	1.006 (0.990-1.022)	0.448
eGFR	-0.029	0.971 (0.887-1.063)	0.528
Serum creatinine	-0.017	0.983 (0.398-2.431)	0.971
Normal Glomeruli	-0.078	0.963 (0.937-0.989)	0.006
Tubular Atrophy and Interstitial Fibrosis	0.386	01472 (0.709-3.058)	0.300
RRT at time of diagnosis	1.603	4.969 (1.675-14.74)	0.004

Ab, antibody; double positive Ab, antibodies for anti-glomerular basement membrane (GBM) and antineutrophil cytoplasmic (ANCA) antibodies; eGFR, estimated glomerular filtration rate; ESKD, end stage kidney disease; HR, Hazard ratio; 95% CI, 95% confidence interval; RRT, renal replacement therapy.

Supplementary Table 2c | Univariable models for time to recovery

	HR (95% CI)	*C*	*P* Value
Ab (Mono Anti-GBM, Double Positive Ab)	1.520 (0.938 - 2.466)	0.579	0.09
Female Sex	1.464 (0.852 - 2.519)	0.539	0.168
Age at diagnosis	1.004 (0.984 - 1.025)	0.529	0.686
eGFR	1.036 (0.969 - 1.107)	0.593	0.297
Normal Glomeruli	1.045 (1.020 - 1.071)	0.622	<0.001
Tubular Atrophy and Interstitial Fibrosis	0.862 (0.291 – 2.549)	0.522	0.799
RRS	0.794 (0.693-0.910)	0.644	<0.001

Ab, antibody; C, Harrell's Concordance; double positive Ab, antibodies for anti-glomerular basement membrane (GBM) and antineutrophil cytoplasmic (ANCA) antibodies; eGFR, estimated glomerular filtration rate; ESKD, end stage kidney disease; HR, Hazard ratio; 95% CI, 95% confidence interval; mono anti-GBM, anti-GBM antibodies without additional ANCA antibodies; RRS, Renal Risk Score comprised of estimated glomerular filtration rate (G0>15 ml/min/1.73 m^2, G1≤15 ml/min/1.73 m^2), percentage of normal glomeruli in the kidney biopsy (N0>25%, N1=10-25%, N2<10%), tubular atrophy and interstitial fibrosis (T0 ≤ mild to moderate, T1 ≥ moderate), with points assigned (G1=3, N1=4, N2=6, T1=2) and risk groups created (low 0, moderate 2 - 7, and high risk group 8 – 11 points).

Supplementary Table 3a | Clinical Outcome based on Percentage of Normal Glomeruli

	N0 (n=33)	N1 (n=30)	N2 (n=111)
Glomeruli on biopsy (Median (IQR))	15.0 (8.0-21.0)	15.0 (9.0-19.8)	17.0 (11.0-25.0)
eGFR (Median ml/min/1.73m^2 (IQR))	17.0 (8.0-34.0)	6.0 (5.0-9.0)	6.0 (4.0-9.0)
Initial need for RRT	11 (33.3%)	23 (76.7%)	95 (85.6%)
Renal Recovery	6/11 (54.5%)	11/23 (47.8%)	16/95 (16.8%)
ESKD	5 (15.2%)	16 (53.3%)	85 (76.6%)
Mortality	5 (15.2%)	11 (36.7%)	37 (33.3%)

eGFR, estimated glomerular filtration rate; ESKD, end stage kidney disease; IQR, interquartile range; N0>25% normal glomeruli; N1=10-25% normal glomeruli; N2<10% normal glomeruli; RRT, renal replacement therapy.

Supplementary Table 3b | Clinical Outcome based on Percentage of Normal Glomeruli in Patients with less than 10% Normal Glomeruli

Normal Glomeruli	>0% (n=22)	0% (n=89)
Glomeruli on biopsy (Median (IQR))	25.5 (21.0-34.0)	14.0 (10.0-22.0)
eGFR (Median ml/min/1.73m^2 (IQR))	5.0 (4.0-9.0)	6.0 (4.0-9.8)
Initial need for RRT	20 (90.9%)	75 (84.3%)
Renal Recovery	3/20 (15.0%)	13/75 (17.3%)
ESKD	17 (77.3%)	68 (76.4%)
Mortality	7 (31.8%)	30 (33.7%)

eGFR, estimated glomerular filtration rate; ESKD, end stage kidney disease; IQR, interquartile range; RRT, renal replacement therapy.