Supplemental Material Table of Contents

Supplemental File 1. Scoring questionnaire

Overall

1 Total number of glomeruli

2 AAGN class

a) Focal

b) Crescentic

c) Mixed

d) Sclerotic

Inflammatory infiltrate present in:

3 Infiltrates

a) <10% of unscarred parenchyma

b) 10 to 25% of unscarred parenchyma

c) 26 to 50% of unscarred parenchyma

d) >50% of unscarred parenchyma

4 Dominant cell type of infiltrate

a) Neutrophils

b) Mononuclear cells

c) Eosinophils

5 Interstitial fibrosis and tubular atrophy

a) No interstitial fibrosis and tubular atrophy

b) Mild interstitial fibrosis and tubular atrophy (<25% of cortical area)

c) Moderate interstitial fibrosis and tubular atrophy (26-50% of cortical area)

d) Severe interstitial fibrosis and tubular atrophy/loss (>50% of cortical area)

6 Intra-epithelial infiltrate

a) No mononuclear cells in tubules

b) Foci with 1 to 4 cells/tubular cross section or 10 tubular cells

c) Foci with 5 to 10 cells/tubular cross section

d) Foci with >10 cells/tubular cross section

Vessels

7 Is vasculitis present in the small vessels (arterioles and/or arteries)?

a) Yes

b) No

8 Are large vessels present in the biopsy?

a) Yes (please answer question 9)

b) No (proceed to question 10)

9 Is vasculitis present in the large vessels?

a) Yes

b) No

Granulomas

10 Are granulomas present?

a) Yes

b) No

Conclusion

11 Do you have any comments?

Supplemental Table 1. The kidney risk score proposed by Brix *et al*.

Risk factor		Points
Percentage normal glomeruli	>25%	0
	10-25%	4
	<10%	6
Tubular atrophy + interstitial fibrosis	≤25%	0
	>25%	2
Kidney function at time of diagnosis (GFR)	>15 ml/min/1.73m^2	0
	≤15 ml/min/1.73m^2	3

Risk group	Points
Low	0
Medium	2-7
High	8-11

Reference: Brix SR, Noriega M, Tennstedt P, Vettorazzi E, Busch M, Nitschke M, Jabs WJ, Ozcan F, Wendt R, Hausberg M, Sellin L, Panzer U, Huber TB, Waldherr R, Hopfer H, Stahl RAK, Wiech T: Development and validation of a renal risk score in ANCA-associated glomerulonephritis. *Kidney Int,* 94**:** 1177-1188, 2018

Supplemental Table 2. Characteristics of the validation cohort

	Total (*n*=145)
Age at biopsy, year, mean±SD	61±13
Male (%)	83 (57)
Diagnosis (%)[a]	
Granulomatosis with polyangiitis	63 (45)
Microscopic polyangiitis	71 (51)
Eosinophilic granulomatosis with polyangiitis	2 (1)
Kidney-limited vasculitis	4 (3)
Diagnostic delay, months, mean±SD	3.1±9.0
ANCA specificity (%)[b]	
PR3	50 (37)
MPO	73 (54)
Negative	6 (4)
Double positive	6 (4)
Center	
Cochin Hospital, Paris	6 (4)
General University Hospital in Prague	38 (26)
Leiden University Medical Center, Leiden	36 (25)
Medical University of Innsbruck	7 (5)
Medical University of Vienna	9 (6)
Necker Hospital, Paris	4 (3)
Rigshospitalet Copenhagen	7 (5)
Teinekeijnkai Hospital Sapporo	7 (5)
JCHO Sendai Hospital, Sendai	24 (17)
Weill Cornell Medical College New York	7 (5)

MPO, myeloperoxidase; PR3, proteinase-3.

[a] The diagnosis was not further specified in 5 patients.

[b] ELISA test results were available in 135 patients.

Supplemental Table 3. Treatment according to histopathological class in the validation cohort

Induction therapy	Total (*n*=143)[a]	Focal class (*n*=51)	Crescentic class (*n*=37)	Mixed class (*n*=39)	Sclerotic class (*n*=16)
Plasma exchange	20 (14)	5 (10)	9 (24)	4 (10)	2 (13)
Corticosteroids only	19 (13)	4 (8)	4 (11)	6 (15)	5 (31)
Corticosteroids and cyclophosphamide	106 (74)	43 (84)	30 (81)	23 (59)	10 (63)
Corticosteroids and azathioprine or MMF	8 (6)	2 (4)	1 (3)	4 (10)	1 (6)
Corticosteroids and mizoribine	5 (4)	1 (2)	0 (0)	4 (10)	0 (0)
Corticosteroids and rituximab[b]	5 (4)	1 (2)	2 (5)	2 (5)	0 (0)

Maintenance therapy	Total (*n*=136)[c]	Focal class (*n*=49)	Crescentic class (*n*=35)	Mixed class (*n*=36)	Sclerotic class (*n*=16)
Initially none	5 (4)	2 (4)	1 (3)	2 (6)	0 (0)
Corticosteroids only	27 (20)	8 (16)	4 (11)	7 (19)	8 (50)
Corticosteroids and cyclophosphamide	8 (6)	5 (10)	1 (3)	2 (6)	0 (0)
Corticosteroids and azathioprine or MMF	83 (61)	29 (59)	25 (71)	21 (58)	8 (50)
Azathioprine or MMF	5 (4)	3 (6)	2 (6)	0 (0)	0 (0)
Corticosteroids and mizoribine	8 (6)	2 (4)	2 (6)	4 (11)	0 (0)

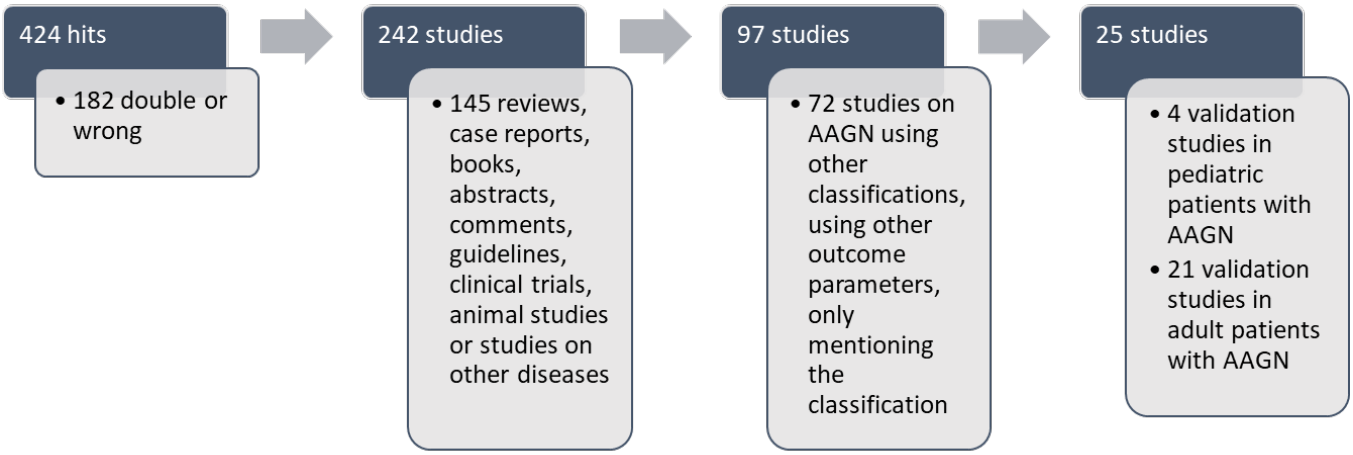
MMF, mycophenolate mofetil.

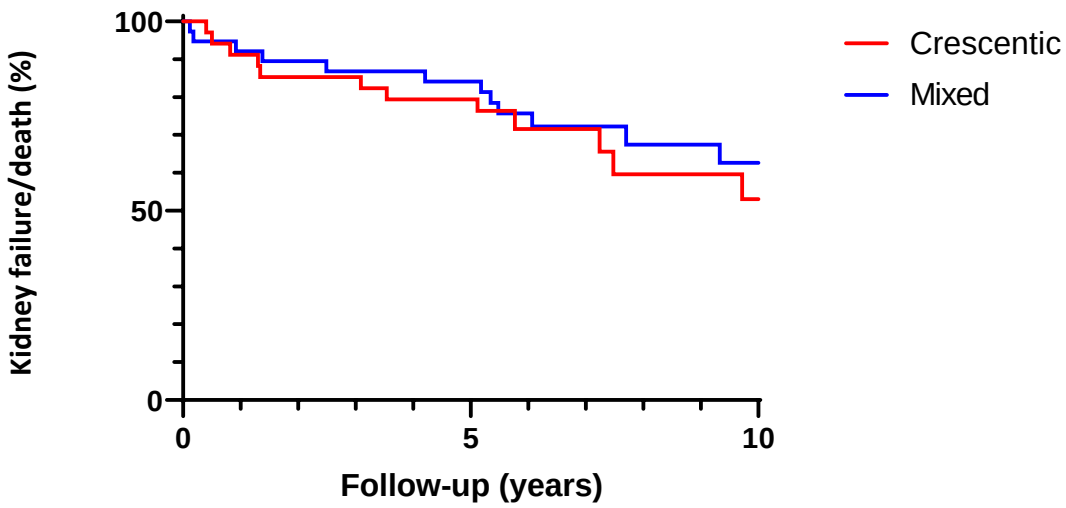
[a] Data on induction therapy was missing in 2 patients.

[b] One of these patients also received 2 doses of intravenous cyclophosphamide.

[c] Data on maintenance therapy was available in 142 patients. Six patients did not receive maintenance therapy due to death or dialysis dependency.

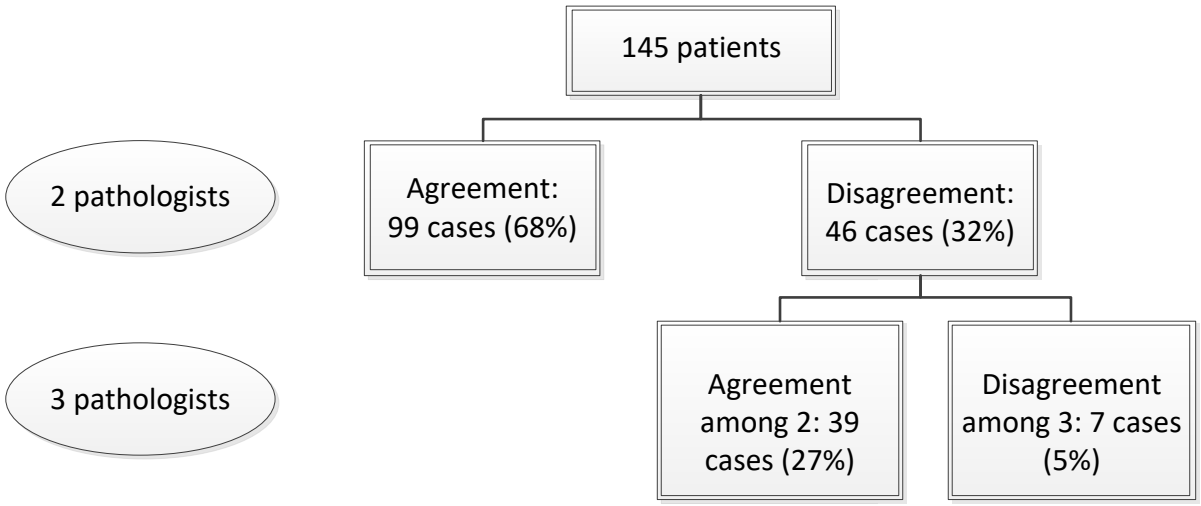
Supplemental Figure 1. Flowchart illustrating how validation studies were selected for the meta-analyses





N at risk						
Crescentic	37	29	27	14	10	7
Mixed	39	34	32	23	14	13

Supplemental Figure 2. Combined outcome of kidney failure/death in the crescentic and mixed classes over time. *P*-value (log-rank) = 0.57



Supplemental Figure 3. Interobserver agreement on histopathological class