

**Supplementary Table S1.** Comparison of chosen parameters between the RI and non- RI groups of all AAV, fulfilling POLVAS definition of respiratory involvement.

	RI group	Non- RI group	<i>p</i> -value
General data			
All AAV	475/620; 76.6%	145/620; 23.4%	-
GPA	315/475; 66.3%	98/145; 67.6%	0.7765
MPA	60/475; 12.6%	45/145; 31.0%	<b>&lt;0.0001</b>
EGPA	100/475; 21.1%	2/145; 1.4%	<b>&lt;0.0001*</b>
Men	238/475; 50.1%	57/145; 39.3%	<b>0.0227</b>
Age at the time of diagnosis (years)	49.8 ± 15.6	52.7 ± 16.3	0.0584
Symptoms and signs <sup>#</sup>			
Cardiovascular system	95/473; 20.1%	13/144; 9.0%	<b>0.0022</b>
Gastrointestinal system	72/473; 15.2%	11/145; 7.6%	<b>0.0183</b>
Central nervous system (CNS)	48/471; 10.2%	6/145; 4.1%	<b>0.0242</b>
Peripheral nervous system	119/464; 25.6%	23/145; 15.9%	<b>0.0150</b>
Deaths and relapses			
Deaths	48/465; 10.3%	8/137; 5.8%	0.1124
Total number of relapses	1.0 (0.0-2.0)	0.0 (0.0-1.0)	<b>0.0035</b>
Relapses requiring hospitalisation (at least one)	227/431; 52.7%	59/141; 41.8%	<b>0.0257</b>
Relapses requiring ICU stay	28/429; 6.5%	2/140; 1.4%	<b>0.0335</b>
Laboratory tests <sup>#</sup>			
p ANCA presence	86/399; 21.6%	42/121; 34.7%	<b>0.0033</b>
ANCA absence	54/399; 13.5%	7/121; 5.8%	<b>0.0203</b>
Eosinophilia	106/417; 25.4%	5/125; 4.0%	<b>&lt;0.0001</b>
Maximal CRP serum concentration (mg/l); <sup>&amp;</sup>	36.0 (11.0-90.0)	25.0 (6.0-75.0)	<b>0.0489</b>
Treatment <sup>#</sup>			
<b>GCs in remission induction</b>	455/473; 96.2%	131/144; 90.1%	<b>0.0120</b>
<b>AZA in maintenance treatment</b>	165/415; 39.8%	42/138; 30.4%	<b>0.0499</b>

The proportions included in the columns described as RI group and non-RI group are the proportions of cases with the relevant feature to all cases with or without RI, respectively.

Data are presented as median and interquartile range or mean and standard deviation, as appropriate.

\*Yates correction used. <sup>&</sup>At the time of diagnosis <sup>#</sup>Only statistically significant values are presented.

PR3: proteinase 3; MPO: myeloperoxidase; GCs: glucocorticosteroids; AZA: azathioprine.

**Supplementary Table S2.** Differences between the RI and the non-RI group in defined subgroups of AAV, fulfilling POLVAS definition of respiratory involvement.

	RI group	Non-RI group	<i>p</i> -value
GPA subgroup			
Men	171/315; 54.3%	37/98; 37.8%	0.0043
Constitutional symptoms	276/314; 87.9%	77/97; 79.4%	0.0352
Renal involvement	206/315; 65.4%	45/96; 46.9%	0.0011
Relapses requiring ICU stay	17/305; 5.6%	0/94; 0.0%	0.0406*
Maximal creatinine concentration (mg/dl); median	1.2 (1.0-3.3)	1.0 (0.9-2.2)	0.0038
Maximal CRP concentration (mg/l); median &	40.0 (10.5-111.0)	22.0 (5.0-75.7)	0.0172
ANCA absence	10/270; 3.7%	7/75; 9.3%	0.0472
Total GCs use <sup>#</sup>	297/313; 94.9%	86/97; 88.7%	0.0307
GCs intravenous use <sup>#</sup>	249/313; 79.6%	60/97; 61.9%	0.0004
GCs without any other immunosuppressive drug <sup>#</sup>	18/313; 5.8%	12/97; 12.4%	0.0287
CYC use <sup>#</sup>	278/313; 88.8%	74/97; 76.3%	0.0020
Plasmaphereses	50/310; 16.1%	6/92; 6.5%	0.0194
GCs pulses use (at least one)	224/279; 80.3%	57/86; 66.3%	0.0070
MTX use <sup>#</sup>	12/313; 3.8%	10/97; 10.3%	0.0134
MTX use <sup>§</sup>	69/308; 22.4%	32/95; 33.7%	0.0265
MPA subgroup			
Relapses requiring hospitalisation	23/60; 38.3%	9/45; 20.0%	0.0434
anti-PR3 presence	3/53; 5.7%	9/42; 21.4%	0.0470*
CYC use <sup>#</sup>	54/60; 90.0%	33/45; 73.3%	0.0249
GPA-MPA subgroup			
Men	205/375; 54.7%	56/143; 39.2%	0.0016
MPA diagnosis	60/375; 16.0%	45/143; 31.5%	0.0001
Central nervous system involvement	40/371; 10.8%	6/143; 4.2%	0.0191
Deaths	47/365; 12.9%	8/135; 5.9%	0.0274
Relapses requiring hospitalisation	205/368; 55.7%	59/139; 42.4%	0.0077
Relapses requiring ICU stay	27/365; 7.4%	2/138; 1.4%	0.0193*
Maximal CRP concentration (mg/l); median &	43.0 (13.0-107.0)	25.0 (8.0-75.0)	0.0118
c ANCA presence	248/327; 75.8%	72/119; 60.5%	0.0015
p ANCA presence	70/327; 21.4%	40/119; 33.6%	0.0082
anti-PR3	256/342; 74.9%	80/129; 62.0%	0.0060
anti-MPO	272/331; 21.8%	41/126; 32.5%	0.0169
GCs intravenous use <sup>#</sup>	302/373; 81.0%	94/142; 66.2%	0.0004
CYC use <sup>#</sup>	332/373; 89.0%	107/142; 75.4%	0.0001
GCs without any other immunosuppressive drug <sup>#</sup>	24/373; 6.4%	22/142; 15.5%	0.0013
Plasmaphereses	61/370; 16.5%	11/137; 8.0%	0.0154
GCs pulses use (at least one)	270/334; 80.8%	86/127; 67.7%	0.0027

Only statistically significant values are presented.

\*Yates correction used. &At the time of diagnosis. <sup>#</sup>In remission induction treatment. <sup>§</sup>In maintenance treatment.