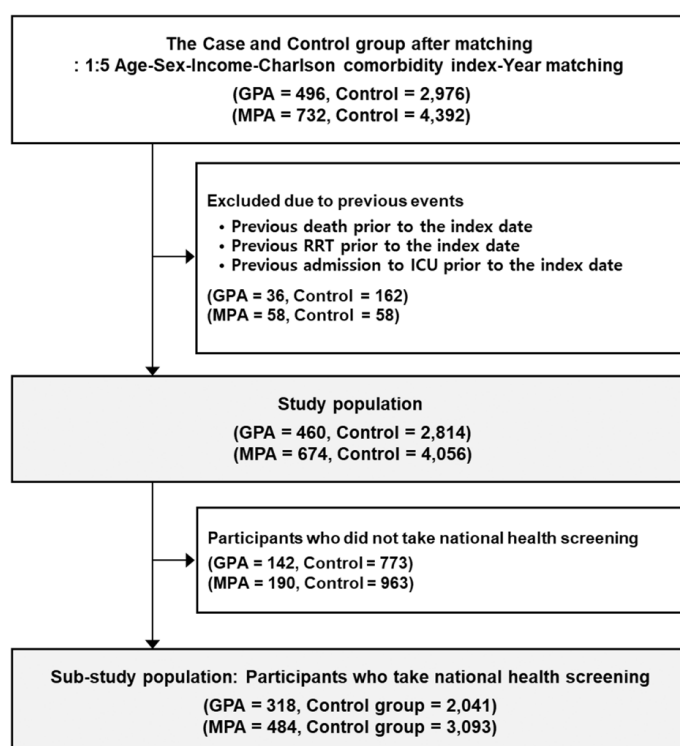
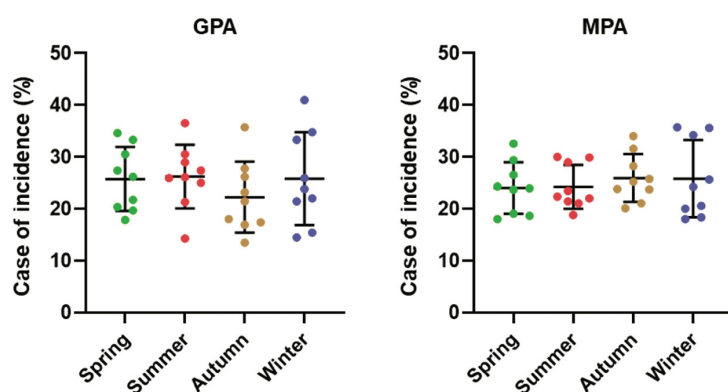


Supplementary Table S1. Diagnosis of patients with only ICD-10 code for GPA and MPA without registration of the RID registry in our institution between 2010 and 2018.

	Patients with ICD-10 code for GPA but no RID registration (n = 38)	Patients with ICD-10 code for MPA but no RID registration (n = 27)
Follow-up loss	2 (5.3%)	3 (11.1%)
Non-rheumatic diseases	21 (55.3%)	12 (44.4%)
Other vasculitis*	3 (7.9%)	9 (33.3%)
Clinically suspected, but no pathologic confirmation of GPA or MPA	10 (26.3%)	2 (22.2%)
True cases of GPA or MPA	2 (5.3%)	1 (11.1%)

GPA: granulomatosis with polyangiitis; ICD-10: 10th revision of the International Statistical Classification of Diseases and Related Health Problems; MPA: microscopic polyangiitis; RID: rare intractable disease.

*including polyarteritis nodosa, immune complex small vessel vasculitis, and eosinophilic granulomatosis with polyangiitis.

**Supplementary Fig. S1.** Flow diagram of study population for patients with granulomatosis with polyangiitis (GPA), microscopic polyangiitis (MPA), and their comparator groups.**Supplementary Fig. S2.** Seasonal incidence of GPA and MPA.

Supplementary Table S2. Annual incidence and prevalence rate (per million) of GPA and MPA between 2010 and 2018 in South Korea.

Incidence rate (per million)	Year	GPA									Total
		2010	2011	2012	2013	2014	2015	2016	2017	2018	
Case, N		52	42	56	69	61	54	59	84	69	546 ^a
Population, N		50,515,666	50,734,284	50,948,272	51,141,463	51,327,916	51,529,338	51,696,216	51,778,544	51,826,059	
Incidence rate		1.03	0.83	1.10	1.35	1.19	1.05	1.14	1.62	1.33	
Incidence rate (per million)	Year	MPA									Total
		2010	2011	2012	2013	2014	2015	2016	2017	2018	
Case, N		42	38	50	59	85	107	117	128	169	795 ^b
Population, N		50,515,666	50,734,284	50,948,272	51,141,463	51,327,916	51,529,338	51,696,216	51,778,544	51,826,059	
Incidence rate		0.83	0.75	0.98	1.15	1.66	2.08	2.26	2.47	3.26	
Prevalence rate (per million)	Year	GPA									Total
		2010	2011	2012	2013	2014	2015	2016	2017	2018	
Case, N		205	229	260	305	337	361	376	421	452	2,946
Population, N		50,515,666	50,734,284	50,948,272	51,141,463	51,327,916	51,529,338	51,696,216	51,778,544	51,826,059	
Prevalence rate		4.06	4.51	5.10	5.96	6.57	7.01	7.27	8.13	8.72	
Prevalence rate (per million)	Year	MPA									Total
		2010	2011	2012	2013	2014	2015	2016	2017	2018	
Case, N		79	81	114	144	202	257	318	390	503	2,088
Population, N		50,515,666	50,734,284	50,948,272	51,141,463	51,327,916	51,529,338	51,696,216	51,778,544	51,826,059	
Incidence rate		1.56	1.60	2.24	2.82	3.94	4.99	6.15	7.53	9.71	

N: number of patients; GPA: granulomatosis with polyangiitis; MPA: microscopic polyangiitis.

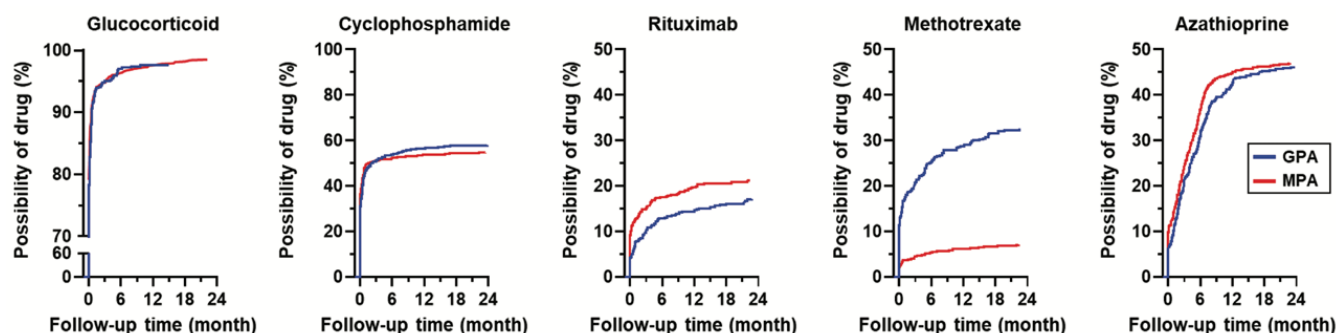
^a10 patients with GPA is excluded, due to missing value of age and sex.

^b8 patients with MPA is excluded, due to missing value of age and sex.

Supplementary Table S3. Age-specific annual incidence rate (per million) of GPA and MPA between 2010 and 2018 in South Korea.

Age	Year	GPA								
		2010	2011	2012	2013	2014	2015	2016	2017	2018
0-19	Case, N	2	0	0	0	1	0	1	12	
	Population, N	11,584,399	11,356,974	11,115,990	10,853,649	10,580,278	10,318,777	10,047,206	9,739,623	9,434,215
	Incidence rate	0.17	0.00	0.00	0.00	0.09	0.00	0.10	0.10	0.21
20-39	Case, N	2	8	3	7	2	5	8	6	3
	Population, N	15,237,505	14,982,202	14,795,891	14,604,943	14,453,891	14,370,014	14,292,568	14,179,616	14,094,116
	Incidence rate	0.13	0.53	0.20	0.48	0.14	0.35	0.56	0.42	0.21
40-59	Case, N	21	14	22	24	26	12	18	2916	
	Population, N	15,911,175	16,374,829	16,627,728	16,945,217	17,146,377	17,183,784	17,221,714	17,192,95	617,104,471
	Incidence rate	1.32	0.85	1.32	1.42	1.52	0.70	1.05	1.69	0.94
60-69	Case, N	16	12	18	24	16	19	16	19	24
	Population, N	4,191,329	4,216,872	4,328,536	4,472,556	4,706,108	5,073,279	5,376,421	5,657,264	5,949,639
	Incidence rate	3.82	2.85	4.16	5.37	3.40	3.75	2.98	3.36	4.03
≥ 70	Case, N	11	8	13	14	16	18	16	2924	
	Population, N	3,591,258	3,803,407	4,080,127	4,265,098	4,441,262	4,583,484	4,758,307	5,009,085	5,243,618
	Incidence rate	3.06	2.10	3.19	3.28	3.60	3.93	3.36	5.79	4.58
Total	Case, N	52	42	56	69	61	54	59	84	69
Age	Year	MPA								
		2010	2011	2012	2013	2014	2015	2016	2017	2018
0-19	Case, N	2	1	1	2	1	2	0	3	1
	Population, N	11,584,399	11,356,974	11,115,990	10,853,649	10,580,278	10,318,777	10,047,206	9,739,623	9,434,215
	Incidence rate	0.17	0.09	0.09	0.18	0.09	0.19	0.00	0.31	0.11
20-39	Case, N	1	3	1	2	3	1	8	3	5
	Population, N	15,237,505	14,982,202	14,795,891	14,604,943	14,453,891	14,370,014	14,292,568	14,179,616	14,094,116
	Incidence rate	0.07	0.20	0.07	0.14	0.21	0.07	0.56	0.21	0.35
40-59	Case, N	11	5	8	12	12	17	20	28	30
	Population, N	15,911,175	16,374,829	16,627,728	16,945,217	17,146,377	17,183,784	17,221,714	17,192,956	17,104,471
	Incidence rate	0.69	0.31	0.48	0.71	0.70	0.99	1.16	1.63	1.75
60-69	Case, N	17	19	19	15	31	30	41	34	51
	Population, N	4,191,329	4,216,872	4,328,536	4,472,556	4,706,108	5,073,279	5,376,421	5,657,264	5,949,639
	Incidence rate	4.06	4.51	4.39	3.35	6.59	5.91	7.63	6.018.57	
≥ 70	Case, N	11	10	21	28	38	57	48	60	82
	Population, N	3,591,258	3,803,407	4,080,127	4,265,098	4,441,262	4,583,484	4,758,307	5,009,085	5,243,618
	Incidence rate	3.06	2.63	5.15	6.56	8.56	12.44	10.09	11.98	15.64
Total	Case, N	42	38	50	59	85	107	117	128	169

N: number of patients; GPA: granulomatosis with polyangiitis; MPA: microscopic polyangiitis.



Supplementary Fig. S3. Cumulative incidence of immunosuppressants prescription in the study population.

Supplementary Table S4. Treatment outcome and prognosis in the subcohort of patients who received national health screening.

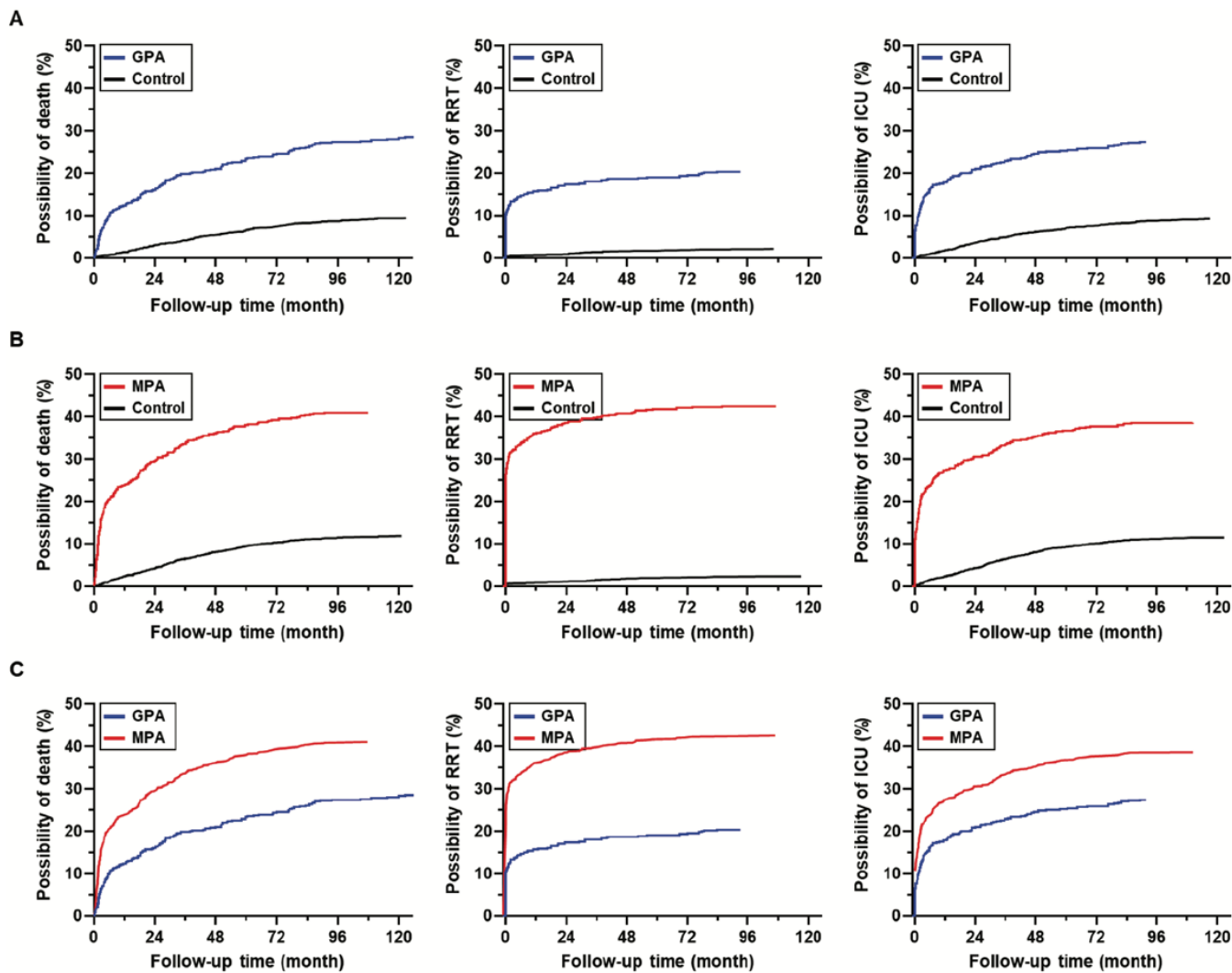
	Study population for GPA		<i>p</i>	Study population for MPA		<i>p</i>
	Matched (N = 1,723)	Patients with GPA (N = 318)		Matched (N = 2,609)	Patients with MPA (N = 484)	
Death						
Event, N (%)	145 (8.4)	90 (28.3)		242 (9.3)	187 (38.6)	
Person-years	10,032	1,532		12,891	1,700	
Unadjusted HR (95% CI)	1.00 (Reference)	4.02 (3.09, 5.24)	<0.001	1.00 (Reference)	5.64 (4.06, 6.83)	<0.001
Adjusted HR (95% CI)						
Model 3	1.00 (Reference)	5.62 (3.48, 9.08)	<0.001	1.00 (Reference)	8.81 (6.65, 11.7)	<0.001
Model 4	1.00 (Reference)	5.39 (3.32, 8.75)	<0.001	1.00 (Reference)	8.60 (6.46, 11.4)	<0.001
Renal replacement therapy						
Event, N (%)	28 (1.63)	67 (21.1)		55 (2.1)	198 (40.9)	
Person-years	9,997	1,378		12,798	1,306	
Unadjusted HR (95% CI)	1.00 (Reference)	15.4 (9.88, 23.9)	<0.001	1.00 (Reference)	26.3 (19.5, 35.5)	<0.001
Adjusted HR (95% CI)						
Model 3	1.00 (Reference)	22.8 (11.9, 43.4)	<0.001	1.00 (Reference)	35.5 (24.2, 52.3)	<0.001
Model 4	1.00 (Reference)	23.7 (12.3, 45.8)	<0.001	1.00 (Reference)	35.9 (24.3, 53.1)	<0.001
ICU admission						
Event, N (%)	144 (8.4)	81 (25.5)		273 (10.5)	184 (38.0)	
Person-years	9,680	1,387		12,350	1,438	
Unadjusted HR (95% CI)	1.00 (Reference)	3.76 (2.87, 4.94)	<0.001	1.00 (Reference)	5.32 (4.41, 6.42)	<0.001
Adjusted HR (95% CI)						
Model 3	1.00 (Reference)	3.91 (2.38, 6.43)	<0.001	1.00 (Reference)	5.88 (4.38, 7.90)	<0.001
Model 4	1.00 (Reference)	4.34 (2.63, 7.24)	<0.001	1.00 (Reference)	5.79 (4.29, 7.81)	<0.001

Hazard ratios were calculated by Cox proportional hazards regression analysis adjusted for following covariates:

Model 3: Model 2 + smoking status, alcohol consumption, and MVPA.

Model 4: Model 3 + body mass index, blood pressure, total cholesterol, and fasting glucose serum.

CI: confidence interval; HR: hazard ratio; ICU: intensive care unit; GPA: granulomatosis with polyangiitis; MPA: microscopic polyangiitis; MVPA: moderate-to-vigorous physical activity.



Supplementary Fig. S4. Kaplan-Meier curves indicating cumulative incidence of all-cause mortality, renal replacement therapy (RRT), and admission to intensive care unit (ICU) compared to the comparator group in patients with GPA (A) and MPA (B). Direct comparison of the incidence of these outcomes showed that all-cause mortality, RRT, and ICU care were more common in patients with MPA (C).