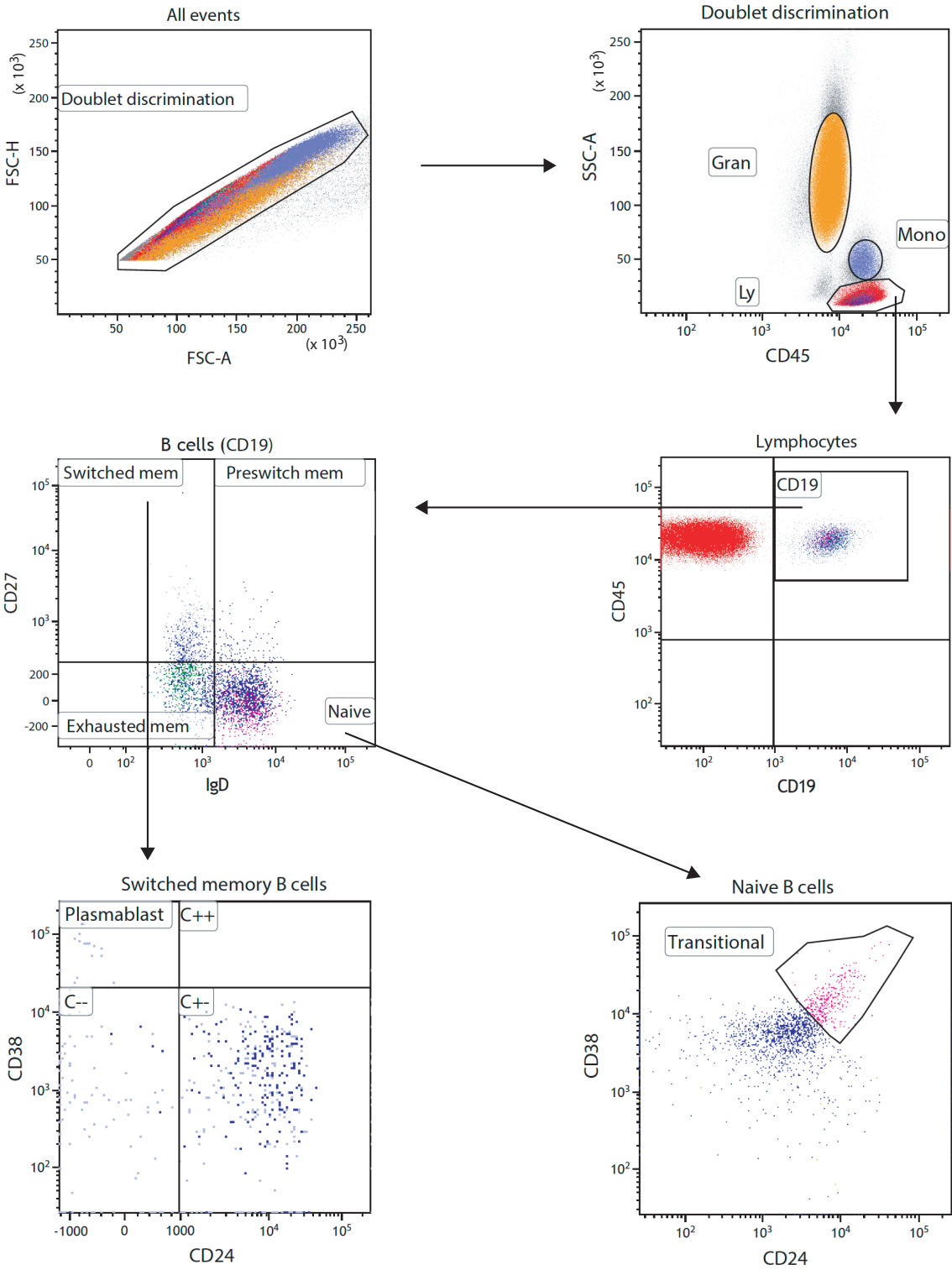
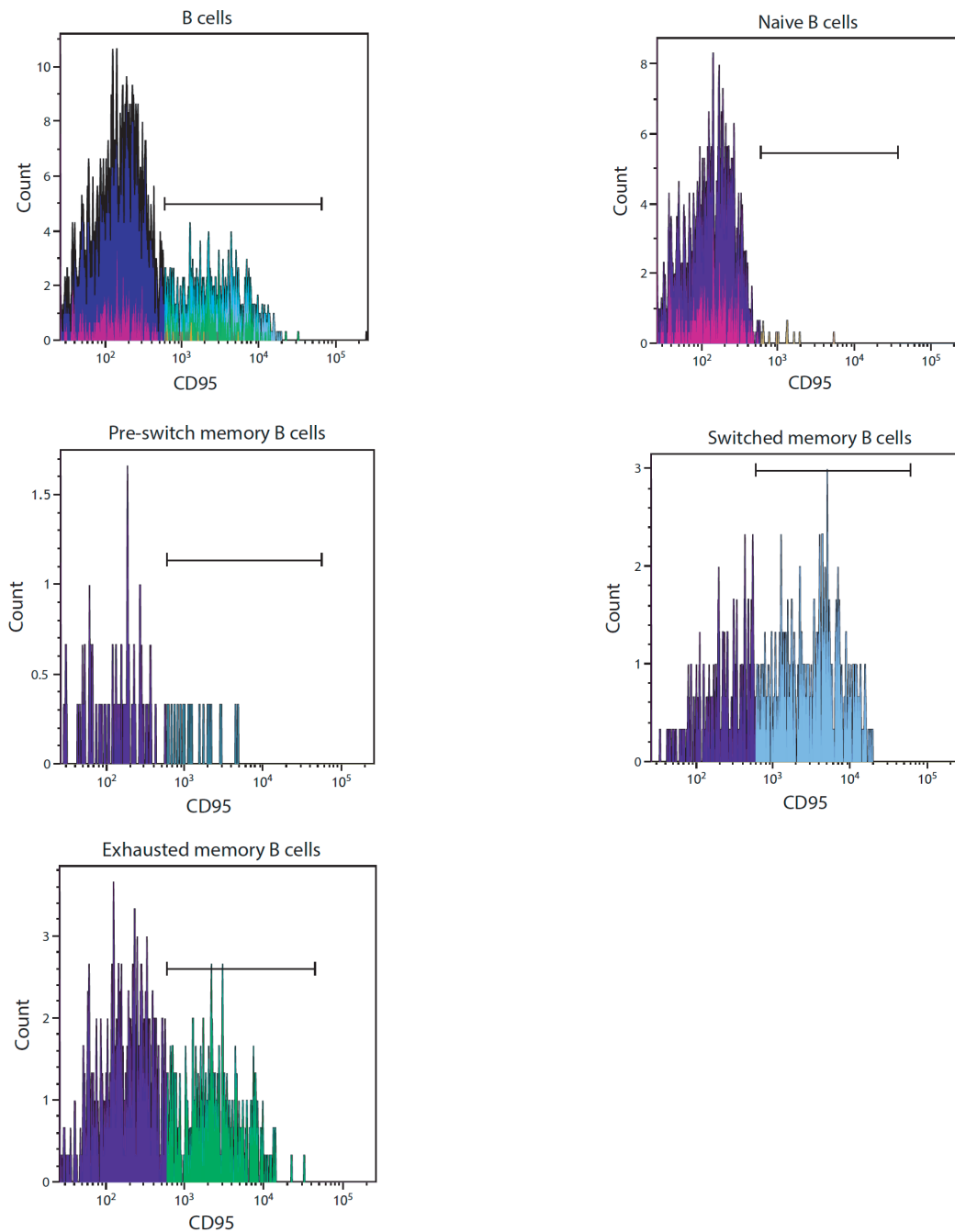


Supplementary Materials





Supplementary Figure 1. Gating strategy for B cell subsets. A gate was first set on all events using the FSC-H/FSC-A plot for doublet discrimination. Then, a gate was set on lymphocytes using the SSC-A/CD45 plot. Within the lymphocytes CD19⁺ B cells were gated using the CD45/CD19 plot. B cells were subsequently divided into naive B cells (CD19⁺CD27⁺IgD⁺), pre-switch memory B cells (CD19⁺CD27⁺IgD⁺), switched memory B cells (CD19⁺CD27⁺IgD⁻) and exhausted memory B cells (CD19⁺CD27⁻IgD⁻) using the CD27/IgD plot. Within the naive B cells, the transitional B cells (CD19⁺CD27⁺IgD⁺CD24⁺CD38⁺⁺) were gated using the CD38/CD24 plot. Within the switched memory B cells, the plasmablasts (CD19⁺CD27⁺IgD⁻CD24⁺CD38⁺⁺) were gated using the CD38/CD24 plot. Activated (CD95⁺) B cells were gated within the respective population using the CD95 plot. The figure shows an example from a patient with anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV).

Supplementary Table 1

Frequencies of B cells and subsets in patients with AAV with regard to medication

Phenotype	AAV with ≥1 drugs (n=80)	AAV no medication (n=26)	HBD (n=134)
BVAS3, median (range)	0 (0-26)	0 (0-21)	
B cells (% of lymphocytes)	4.29 (2.65-6.74) ^{a,b}	9.36 (7.39-13.7)	8.33 (5.98-11.6)
B cells (10⁶/L)*	21.5 (10.3-45.0) ^a	86.1 (40.3-153)	
B cell subsets (% of B cells)			
Naive	44.6 (27.9-56.4) ^{a,b}	65.3 (51.2-71.9)	56.7 (47.9-67.0)
Pre-switch memory	7.21 (4.40-10.7) ^a	4.29 (2.66-6.35) ^c	7.00 (4.65-10.2)
Switched memory	24.3 (17.3-34.4) ^{a,b}	9.94 (6.78-16.8)	16.5 (11.4-21.8)
Exhausted memory	21.1 (14.9-25.6) ^b	21.6 (11.4-26.2)	17.4 (12.0-21.6)
Transitional	0.235 (0.00500-1.74) ^{a,b}	3.65 (1.61-6.10)	4.44 (2.83-6.18)
Plasmablasts	1.00 (0.468-2.49) ^b	0.565 (0.348-1.13) ^c	0.320 (0.160-0.613)
Activated B cells and subsets			
CD95⁺ B cells (% of B cells)	25.6 (14.9-35.7) ^{a,b}	12.3 (6.88-17.1)	10.3 (7.24-15.5)
CD95⁺ naive (% of naive)	4.75 (1.98-9.38) ^{a,b}	1.16 (0.518-2.86)	1.02 (0.575-1.71)
CD95⁺ pre-switch memory (% of pre-switch memory)	31.1 (16.8-41.1) ^{a,b}	17.1 (9.93-28.9)	13.5 (9.07-19.8)
CD95⁺ switched memory (% of switched memory)	60.8 (52.2-72.9) ^b	56.4 (40.7-77.7) ^c	44.6 (36.0-51.9)
CD95⁺ exhausted memory (% of exhausted memory)	27.8 (18.0-38.6) ^{a,b}	16.1 (9.13-25.4)	14.0 (8.38-18.2)

Frequencies of B cells (CD19⁺ lymphocytes) and subsets analysed in patients and healthy controls using flow cytometry. The patients were either medicated with ≥1 of the following medications; prednisolone, azathioprine, methotrexate, mycophenolate mofetil, cyclophosphamide, or none of them. Kruskal-Wallis with Dunn's multiple comparisons test was used to calculate level of significance. Mann-Whitney test was used to calculate level of significance for absolute values. Data are presented with medians and interquartile ranges. AAV anti-neutrophil cytoplasmic autoantibody (ANCA)-associated vasculitis, HBD healthy blood donors, BVAS3 Birmingham Vasculitis Activity Score version 3. ^aDenotes significant difference between "AAV with ≥1" drugs and "AAV no medication". ^bDenotes significant difference between "AAV with ≥1" drugs and HBD. ^cDenotes significant difference between "AAV no medication" and HBD. *n equals 76 (AAV with ≥1 drugs), 25 (AAV no medication) for absolute values.