

Supplemental Materials.

Supplemental Table 1. Primer and Probes for TaqMan

Supplemental Table 2. HLADPB1*04:01 in patient population and local healthy controls.

Supplemental Figure 1. Gating schemes for flow cytometry staining. A) gating scheme for detection of peptide bound HLADPB+ cells shown with no peptide control. B) Example gating on a patient with PR3-ANCA homozygous for 04:01 prior to expansion C) Gating scheme for detection of tetramer positive cells post expansion.

Supplemental Figure 2. The amino acid sequence alignment between DPB1*04:01 and DPB1*04:02 shows differences at position 84, 85 and position 65 (A>V is conserved, likely with minimal impact on peptide binding).

Supplemental Figure 3. HLA-DPB1 mRNA and protein expression do not differ between patients and healthy controls with DPB1. A) mRNA expression for DPB1 between disease states of PR3-ANCA patients (all DPB1*04:01 carriers) and healthy controls. No statistical differences between patients with active disease, remitting disease and healthy control carriers or patient non-carriers (Mann-Whitney test with a Bonferroni correlation). HLA-DPB1 gene expression is not dependent on allele copy number (denoted by shape). B) Mean fluorescence intensity (MFI) of DPB1 by flow cytometry on DPB1+ PBMCs to assess protein expression.

Supplemental Figure 4. Phenotype of peptide stimulated T cells. Ex vivo cryopreserved PBMCs from PR3-ANCA patients (active and LTROT) or healthy controls (HC) were incubated with PR3₂₂₅₋₂₃₉, scrambled peptide or no peptide. Cell supernatant was analyzed for 6a) IL10 levels were reduced in the active patient populations, 6b) IFN γ was increased in response to PR3₂₂₅₋₂₃₉ only in the active PR3-ANCA cohort and 6c) surface expression of CCR6 on HLA-DPB tetramer positive cells was lower in the LTROT cohort.

Supplemental Figure 5. HLA-DRB1*04:01 loaded tetramers were used as a control and did not identify autoreactive T cell population.

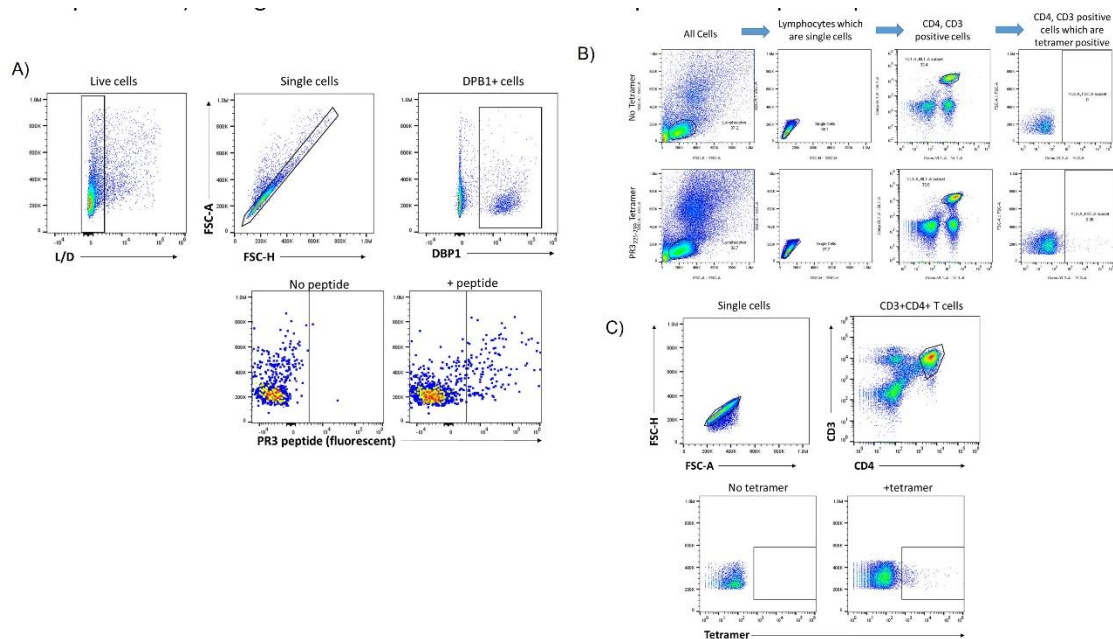
Supplemental Table 1. Primer and Probes for TaqMan

Primers and Probe for TaqMan quantitative real-time PCR			
Gene	Primer	Sequence	Company
COX5B	Forward	5'-TGG CAT CTG GAG GTG TT-3'	Integrated DNA Technologies, INC Coralville, IA
	Reverse	5'-GTC CAG TCC CTT CTT TGC AGC-3'	
	Probe	FAM-TGA TGA AGA GCA GGC GAC TGG GTT G-MGB	
Gene	TaqMan Gene Expression Assay		Company
HLADPB1	Hs485130		ThermoFisher, Waltham, MA
COX5B	HS00426948_m1		

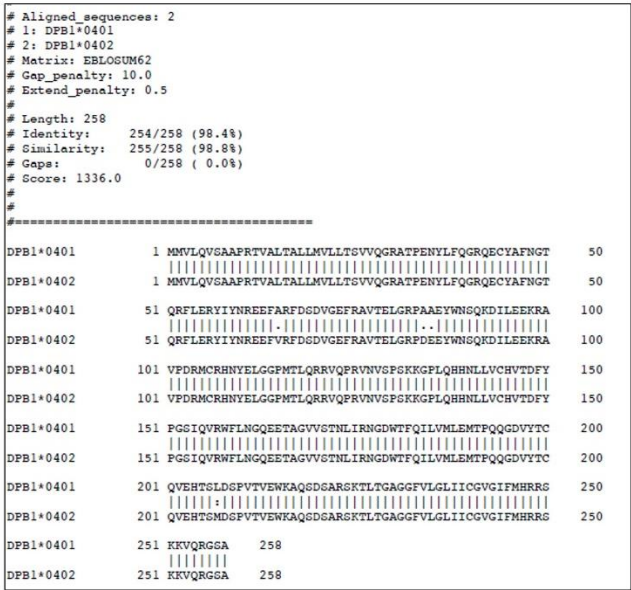
Supplemental Table 2. HLADPB1*04:01 in patient population and local healthy controls.

	Non-carrier (n)	Heterozygous carriers (n)	Homozygous carriers (n)	Total (n)	Carriers total (%)	DPB1*04:01 alleles	Allele Frequency
MPO-ANCA cohort	57	80	30	167	66%	140	0.42
PR3-ANCA cohort	27	55	71	153	82%	197	0.64
Total UNC ANCA Cohort	84	135	101	320	74%	337	0.53
Local Healthy Control Cohort	50	41	13	104	52%	67	0.32

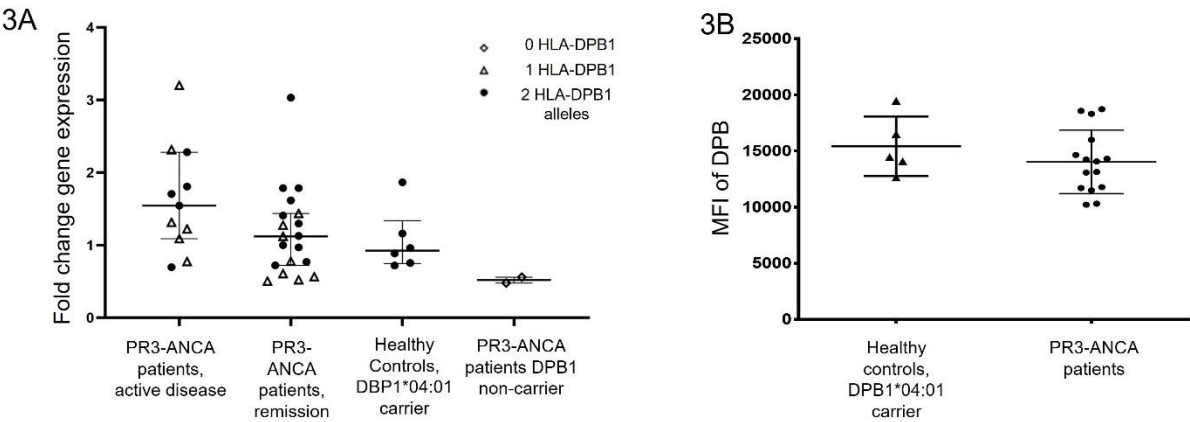
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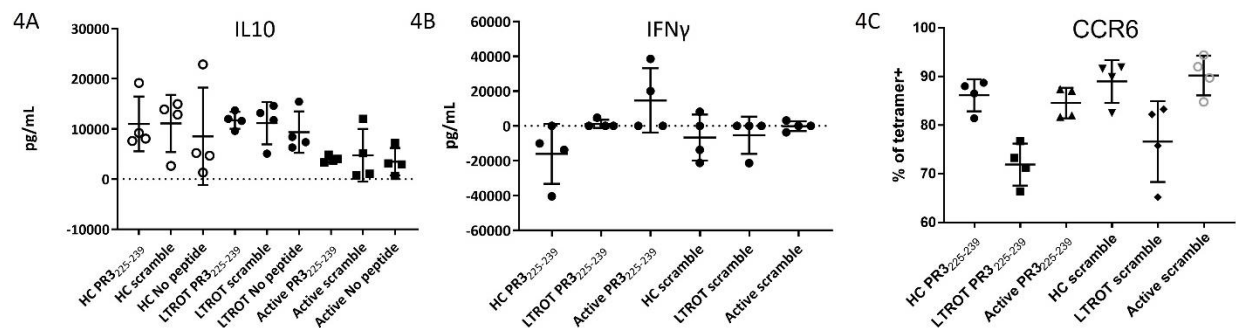
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