

**Article title:** HLA-DQ and HLA-DRB1 alleles associated with Henoch-Schönlein purpura nephritis in Finnish pediatric population: A genome-wide association study

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**Table S1.** Polymorphisms associated to HSP susceptibility in GWAS at the genome-wide significance level  $p < 5 \times 10^{-08}$

SNP	Position	Minor allele	p-value	OR [95% CI]	Frequency HSP	Frequency reference population
rs7754768	32.452.402	C	6.57e-10	0.14 [0.10-0.20]	0.14	0.49
rs9268832	32.460.012	T	6.57e-10	0.14 [0.10-0.20]	0.14	0.49
rs9275578	32.711.607	T	1.06e-09	4.62 [3.51-6.10]	0.71	0.31
rs9275582	32.712.293	T	1.46e-09	4.54 [3.45-5.98]	0.71	0.31
rs2213585	32.445.373	G	2.71e-09	0.15 [0.11-0.22]	0.14	0.48
rs2213586	32.445.317	A	2.71e-09	0.15 [0.11-0.22]	0.14	0.49
rs2227139	32.445.682	G	2.71e-09	0.15 [0.11-0.22]	0.14	0.49
rs3763327	32.446.053	G	2.71e-09	0.15 [0.11-0.22]	0.14	0.49
rs7192	32.443.869	T	2.71e-09	0.15 [0.11-0.22]	0.14	0.49
rs7195	32.444.762	A	2.71e-09	0.15 [0.11-0.22]	0.14	0.49
rs3998158	32.714.215	C	3.97e-09	4.39 [3.33-5.79]	0.70	0.30
rs112396255	32.623.692	G	5.35e-09	4.50 [3.42-5.91]	0.58	0.22
rs17533090	32.622.945	T	5.35e-09	4.50 [3.42-5.91]	0.58	0.22
rs13194877	32.687.771	A	6.06e-09	4.48 [3.41-5.89]	0.57	0.20
rs9275572	32.711.222	A	7.58e-09	0.17 [0.12-0.24]	0.13	0.48
rs17202717	32.430.086	T	8.59e-09	4.33 [3.31-5.67]	0.57	0.20
rs17496307	32.433.259	T	8.59e-09	4.33 [3.31-5.67]	0.57	0.20
rs17496549	32.441.931	T	8.59e-09	4.33 [3.31-5.67]	0.57	0.20
rs17212483	32.697.473	T	8.78e-09	4.27 [3.27-5.58]	0.57	0.20
rs115255544	32.696.851	G	1.14e-08	4.23 [3.24-5.52]	0.57	0.20
rs17212462	32.697.348	T	1.14e-08	4.23 [3.24-5.52]	0.57	0.20
rs17212553	32.697.910	T	1.14e-08	4.23 [3.24-5.52]	0.57	0.20
rs17427445	32.695.987	A	1.14e-08	4.23 [3.24-5.52]	0.57	0.20
rs17427564	32.699.290	C	1.14e-08	4.23 [3.24-5.52]	0.57	0.20
rs34492195	32.703.056	A	1.14e-08	4.23 [3.24-5.52]	0.57	0.20
rs34677551	32.696.159	C	1.14e-08	4.23 [3.24-5.52]	0.57	0.20
rs35472814	32.692.120	C	1.14e-08	4.23 [3.24-5.52]	0.57	0.20
rs35900855	32.696.261	G	1.14e-08	4.23 [3.24-5.52]	0.57	0.20
rs9275516	32.706.866	A	1.14e-08	0.18 [0.13-0.25]	0.13	0.48

rs10484561	32.697.643	G	1.67e-08	4.22 [3.22-5.52]	0.57	0.20
rs35332745	32.706.448	C	1.67e-08	4.11 [3.16-5.35]	0.57	0.20
rs12529093	32.448.081	G	1.91e-08	4.13 [3.16-5.38]	0.58	0.21
rs13209059	32.448.301	G	1.91e-08	4.13 [3.16-5.38]	0.58	0.21
rs13209234	32.448.198	A	1.91e-08	4.13 [3.16-5.38]	0.58	0.21
rs13220657	32.456.254	T	1.91e-08	4.13 [3.16-5.38]	0.58	0.21
rs28895028	32.447.434	A	1.91e-08	4.13 [3.16-5.38]	0.58	0.21
rs28895205	32.457.527	G	1.91e-08	4.13 [3.16-5.38]	0.58	0.21
rs28732229	32.459.604	T	1.91e-08	4.13 [3.16-5.38]	0.58	0.21
rs28895136	32.453.727	C	1.91e-08	4.13 [3.16-5.38]	0.58	0.21
rs3177928	32.444.658	A	1.91e-08	4.13 [3.16-5.38]	0.58	0.21
rs114169440	32.695.144	A	1.99e-08	4.08 [3.13-5.31]	0.57	0.20
rs17599077	32.623.281	C	2.25e-08	4.08 [3.13-5.32]	0.54	0.19
rs4530903	32.614.112	T	2.25e-08	4.08 [3.13-5.32]	0.54	0.19
rs6932517	32.710.405	C	2.41e-08	0.19 [0.14-0.27]	0.13	0.48
rs9275580	32.711.685	G	2.73e-08	3.96 [3.03-5.19]	0.72	0.33
rs2858331	32.713.500	G	3.74e-08	3.82 [2.93-4.97]	0.65	0.33
rs113585425	32.465.348	C	3.83e-08	4.04 [3.10-5.28]	0.53	0.19
rs72847996	32.417.115	A	3.83e-08	4.04 [3.10-5.28]	0.53	0.19

**Table S2.** HLA alleles associated with HSP and IBD with a significance level of  $p < 0.05$ 

HLA allele	HSP (n = 46)			IBD (n = 49)			Reference population (n = 18 757)
	p-value	OR [95% CI]	Allele frequency	p-value	OR [95% CI]	Allele frequency	Allele frequency
DQB1*05:01	4.99 x 10 <sup>-09</sup>	4.37 [3.35 – 5.71]	0.58	0.0016	0.36 [0.26 – 0.51]	0.11	0.2
DQA1*0101	1.04 x 10 <sup>-08</sup>	4.18 [3.21 – 5.45]	0.55	0.0014	0.35 [0.24 – 0.49]	0.10	0.19
DRB1*01:01	2.37 x 10 <sup>-08</sup>	4.10 [3.14 – 5.35]	0.54	0.0013	0.34 [0.24 – 0.49]	0.10	0.19
DQA1*05:01	0.0013	0.04 [0.01 – 0.18]	0	-	-	-	0.10
C*04:01	0.0018	2.21 [1.70 – 2.87]	0.35	-	-	-	0.16
DQB1*02:01	0.0020	0.05 [0.01 – 0.20]	0	-	-	-	0.10
DRB1*03:01	0.0020	0.05 [0.01 – 0.20]	0	-	-	-	0.10
B*35:01	0.0032	2.15 [1.64 – 2.81]	0.30	0.039	0.50 [0.36 – 0.72]	0.10	0.13
DPB1*04:02	0.0053	2.13 [1.61 – 2.81]	0.37	-	-	-	0.20
DQA1*04:01	0.0056	0.21 [0.11 – 0.40]	0.02	-	-	-	0.09
DQB1*04:02	0.0073	0.22 [0.12 – 0.43]	0.02	-	-	-	0.09
DQA1*01:02	0.0080	0.33 [0.21 – 0.52]	0.07	-	-	-	0.19
DRB1*08:01	0.0093	0.27 [0.15 – 0.48]	0.03	-	-	-	0.09
C*12:03	0.015	6.73 [2.85 – 15.88]	0.04	0.00036	12.27 [5.75 – 26.21]	0.06	0.04
DPB1*01:01	0.018	0.09 [0.02 – 0.36]	0	-	-	-	0.06
DRB1*13:01	0.019	0.29 [0.16 – 0.53]	0.03	-	-	-	0.10
A*25:01	0.022	7.84 [2.66 – 23.1]	0.03	0.017	12.67 [4.15 – 38.73]	0.02	0.01
B*08:01	0.024	0.27 [0.14 – 0.52]	0.02	-	-	-	0.09
DRB1*04:04	0.025	3.12 [1.84 – 5.27]	0.09	-	-	-	0.04
A*02:01	0.033	0.56 [0.43 – 0.74]	0.25	-	-	-	0.33
C*07:02	0.034	0.44 [0.29 – 0.67]	0.08	-	-	-	0.15
B*44:02	0.038	2.51 [1.57 – 4.00]	0.10	-	-	-	0.06
DQB1*06:02	0.042	0.42 [0.27 – 0.66]	0.07	-	-	-	0.15

DQA1*01:03	0.044	0.36 [0.2 – 0.63]	0.03	-	-	-	0.09
DQB1*06:03	0.044	0.36 [0.2 – 0.63]	0.03	-	-	-	0.09
A*32:01	-	-	-	0.00089	5.5 [3.18 – 9.50]	0.08	0.03
B*39:01	-	-	-	0.0081	3.92 [2.27 – 6.76]	0.07	0.04
C*02:02	-	-	-	0.0098	3.06 [1.95 – 4.78]	0.10	0.06
DPB1*17:01	-	-	-	0.020	94.59 [23.61 – 378.9]	0.02	0

**Table S3.** Power analysis results for top genome-wide significant SNPs and HLA alleles in HSP

<b>SNP or HLA allele</b>	<b>Power</b>
rs7754768	0.92
rs9275578	0.99
DQA1*01:01	0.97
DQB1*05:01	0.98
DRB1*01:01	0.96

**Table S4.** HSP data according to DQB105:01/DQA101:01/DRB1\*01:01 haplotype

	Other haplotypes	DQB1*05:01/DQA1*01:01 /DRB1*01:01	p-value
Number of patients	26	20	
Gender (male:female)	10:16	9:11	0.77
Renal involvement, n (%)	25 (96)	19 (95)	
At the renal biopsy <sup>a</sup>			
Age (years)	9.8 ± 3.6	9.8 ± 2.7	0.98
Plasma creatinine (µmol/L)	51 ± 21	51 ± 15	0.81
eGFR (mL/min/1.73m <sup>2</sup> )	110 ± 28	107 ± 27	0.48
dU-Prot (g/day)	3.2 (1.6 – 6.3)	1.9 (0.9 – 4.9)	0.36
dU-Prot (mg/h/m <sup>2</sup> )	105 (66 – 242)	83 (31 – 176)	0.38
Nephrotic-range proteinuria, n (%)	18 (78)	12 (63)	0.32
Plasma albumin (g/L)	30.4 ± 8.9	29.2 ± 6.2	0.39
ISKDC ≥ III, n (%)	17 (74)	17 (89)	0.26
At last assessment <sup>b</sup>			
Proteinuria <sup>c</sup> , n (%)	5 (20)	7 (37)	0.31
Follow-up (years)	8.3 (5.4 – 13.6)	9.9 (6.9 – 15.6)	0.36

<sup>a</sup>n=42<sup>b</sup>n=44 (includes patients with HSN; two patients without renal involvement were excluded)<sup>c</sup>Defined as urine protein to creatinine ratio > 20 mg/mmol or positive dipstick test (1+ to 3+)

**Table S5.** HSP data according to B35:01/DQB105:01/DQA101:01/DRB1\*01:01 haplotype

	Other haplotypes	B*35:01/DQB1*05:01/ DQA1*01:01/DRB1*01:01	p-value
Number of patients	32	14	
Gender (male:female)	14:18	5:9	0.75
Renal involvement, n (%)	31 (97)	13 (93)	
At the renal biopsy <sup>a</sup>			
Age (years)	9.8 ± 3.4	9.9 ± 2.8	0.91
Plasma creatinine (µmol/L)	50 (40 – 54)	44 (40 – 65)	0.91
eGFR (mL/min/1.73m <sup>2</sup> )	109 ± 26	107 ± 31	0.81
dU-Prot (g/day)	3.2 (1.0 – 6.6)	1.9 (1.3 – 4.8)	0.60
dU-Prot (mg/h/m <sup>2</sup> )	104 (40 – 250)	83 (37 – 162)	0.54
Nephrotic-range proteinuria, n (%)	21 (72)	9 (69)	>0.99
Plasma albumin (g/L)	30.2 ± 9.0	29.2 ± 4.4	0.43
ISKDC ≥ III, n (%)	22 (76)	12 (92)	0.40
At last assessment <sup>b</sup>			
Proteinuria <sup>c</sup> , n (%)	6 (19)	6 (46)	0.14
Follow-up (years)	7.7 (4.5 – 13.6)	12.0 (8.0 – 16.1)	0.11

<sup>a</sup>n=42<sup>b</sup>n=44 (includes patients with HSN; two patients without renal involvement were excluded)<sup>c</sup>Defined as urine protein to creatinine ratio > 20 mg/mmol or positive dipstick test (1+ to 3+)

**Table S6.** Common SNPs within the HLA region in genetic correlation analysis between HSP and IBD

<b>SNP</b>	<b>Gene</b>	<b>Gene description</b>	<b>Ensembl gene id</b>	<b>Chromosome</b>	<b>Start position</b>	<b>End position</b>
rs115628514	HLA-B	major histocompatibility complex, class I, B	ENSG00000234745	6	31269491	31357188
rs4530903	HLA-DRB1	major histocompatibility complex, class II, DR beta 1	ENSG00000196126	6	32578769	32589848
rs9271344	HLA-DRB1	major histocompatibility complex, class II, DR beta 1	ENSG00000196126	6	32578769	32589848
rs35472814	HLA-DQB1	major histocompatibility complex, class II, DQ beta 1	ENSG00000179344	6	32659467	32668383
rs114169440	HLA-DQB1	major histocompatibility complex, class II, DQ beta 1	ENSG00000179344	6	32659467	32668383
rs35122881	HLA-DQB1	major histocompatibility complex, class II, DQ beta 1	ENSG00000179344	6	32659467	32668383
rs17427445	HLA-DQB1	major histocompatibility complex, class II, DQ beta 1	ENSG00000179344	6	32659467	32668383
rs34677551	HLA-DQB1	major histocompatibility complex, class II, DQ beta 1	ENSG00000179344	6	32659467	32668383
rs35900855	HLA-DQB1	major histocompatibility complex, class II, DQ beta 1	ENSG00000179344	6	32659467	32668383
rs115255544	HLA-DQB1	major histocompatibility complex, class II, DQ beta 1	ENSG00000179344	6	32659467	32668383
rs17212462	HLA-DQB1	major histocompatibility complex, class II, DQ beta 1	ENSG00000179344	6	32659467	32668383
rs17212483	HLA-DQB1	major histocompatibility complex, class II, DQ beta 1	ENSG00000179344	6	32659467	32668383
rs10484561	HLA-DQB1	major histocompatibility complex, class II, DQ beta 1	ENSG00000179344	6	32659467	32668383
rs17212553	HLA-DQB1	major histocompatibility complex, class II, DQ beta 1	ENSG00000179344	6	32659467	32668383
rs17427564	HLA-DQB1	major histocompatibility complex, class II, DQ beta 1	ENSG00000179344	6	32659467	32668383