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On the pathogenesis and clinical outcome of ANCA-associated vasculitis
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Supplementary table 1. Literature search strategies

Database	Search Strategy
PubMed	<p><i>Two strategies:</i></p> <p><u>L. AAV and genes</u> (“Anti-Neutrophil Cytoplasmic Antibody-Associated Vasculitis”[Mesh] OR “ANCA-associated vasculitis”[all fields] OR “ANCA associated vasculitis”[all fields] OR “Anti-Neutrophil Cytoplasmic Antibody-Associated Vasculitis”[all fields] OR “Pauci-Immune Vasculitis”[all fields] OR “Pauci Immune Vasculitis”[all fields] OR “Pauci-Immune Vasculitides”[all fields] OR “ANCA-Associated Vasculitides”[all fields] OR “ANCA Associated Vasculitides”[all fields] OR “Pauci-Immune Vasculitides”[all fields] OR “ANCA-Associated Vasculitides”[all fields] OR “ANCA Associated Vasculitides”[all fields] AND vasculit*[all fields] OR (“Anti-Neutrophil”[all fields] AND “Cytoplasmic”[all fields] AND “Antibody-Associated”[all fields] AND vasculit*[all fields]) OR “Churg-Strauss Syndrome”[all fields] OR OR “Churg Strauss Syndrome”[all fields] OR “Allergic Granulomatous Angitis”[all fields] OR “Allergic Granulomatous Angitides”[all fields] OR “Allergic Angritis”[all fields] OR “Churg-Strauss Vasculitis”[all fields] OR “eosinophilic granulomatosis with polyangitis”[all fields] OR (“cosinophilic”[all fields] AND “granulomatosis”[all fields] AND “polyangitis”[all fields]) OR “Microscopic Polyangitis”[all fields] OR “Wegener’s Granulomatosis”[all fields] OR “Wegeners Granulomatosis”[all fields] OR “Wegener Granulomatosis”[all fields] OR “granulomatosis with polyangitis”[all fields] AND “polyangitis”[all fields] OR “anca”[tw] OR (“pr3”[all fields] OR “pr-3”[all fields] OR “mpo”[all fields]) AND (“Vasculitis”[mesh] OR vasculit*[all fields] OR “anca”[tw])) OR “Antibodies, Antineutrophil Cytoplasmic”[Mesh] OR “Antineutrophil Cytoplasmic Antibody”[all fields] OR “Anti-Neutrophil Cytoplasmic Antibody”[all fields] OR “Anti Neutrophil Cytoplasmic Antibodies”[all fields] OR “Anti-Neutrophil Cytoplasmic Antibodies”[all fields] AND (“genes”[mesh] OR “genes”[all fields] OR “gene”[all fields] OR “genetic”[all fields] OR “Antineutrophil Cytoplasmic Antibodies”[all fields]) AND (“genes”[mesh] OR “genes”[all fields] OR “gene”[all fields] OR “genetic”[all fields] OR “genetics”[Subheading] OR “genetics”[all fields] OR “genetics”[mesh] OR “polymorphism, genetic”[mesh] OR “polymorphism”[all fields] OR “polymorphisms”[all fields] OR “dna”[mesh] OR “dna”[all fields] OR “genome”[mesh] OR “genome”[all fields] OR “genomes”[all fields] OR “genomics”[mesh] OR “genomics”[all fields] OR “genomic”[all fields] OR “Genetic Phenomena”[mesh] OR “Genetic Structures”[Mesh])</p>

Supplementary table 1. Literature search strategies (Continued)

Database	Search Strategy
PubMed	<p>2. Vasculitis and genes (excluding AAV) ((“Vasculitis”[majr] OR vasculit*[ti]) AND (“genes”[majr] OR “gene”[ti] OR “genetic”[ti] OR “genetics”[ti] OR “genetics”[majr] OR “polymorphism, genetic”[majr] OR “polymorphisms”[ti] OR “dna”[majr] OR “dna”[ti] OR “genome”[majr] OR “genome”[ti] OR “genomes”[ti] OR “genomics”[majr] OR “genomic”[ti] OR “Genetic Phenomena”[majr] OR “Genetic Structures”[majr])) NOT ((“Anti-Neutrophil Cytoplasmic Antibody-Associated Vasculitis”[Mesh] OR “ANCA-associated vasculitis”[all fields] OR “ANCA associated vasculitis”[all fields] OR “Anti-Neutrophil Cytoplasmic Antibody-Associated Vasculitis”[all fields] OR “Pauci-Immune Vasculitis”[all fields] OR “Pauci Immune Vasculitis”[all fields] OR “ANCA-Associated Vasculitides”[all fields] OR “ANCA-Associated Vasculitides”[all fields] OR “ANCA Associated Vasculitides”[all fields] AND “Antibody-Associated”[all fields] AND vasculit*[all fields] OR (“Anti-Neutrophil”[all fields] AND “Cytoplasmic”[all fields] AND “Antibody-Associated”[all fields] AND vasculit*[all fields] OR “Churg-Strauss Syndrome”[all fields] OR “Allergic Granulomatous Angiitis”[all fields] OR “Allergic Granulomatous Angiitides”[all fields] OR “Churg-Strauss Vasculitis”[all fields] OR “eosinophilic granulomatosis with polyangiitis”[all fields] OR (“eosinophilic”[all fields] AND “granulomatosis”[all fields] AND “polyangiitis”[all fields] OR “Microscopic Polyangiitis”[all fields] OR “Wegener’s Granulomatosis”[all fields] OR “Wegeners Granulomatosis”[all fields] OR “granulomatosis with polyangiitis”[all fields] AND “polyangiitis”[all fields] OR “anca”[tw]) OR “Antibodies, Antineutrophil Cytoplasmic”[Mesh] OR “Antineutrophil Cytoplasmic Antibody”[all fields] OR “ANCA”[all fields] OR “Antibodies, Antineutrophil Cytoplasmic”[all fields] OR “Anti-Neutrophil Cytoplasmic Antibodies”[all fields] OR “Anti Neutrophil Cytoplasmic Antibodies”[all fields] OR “Antineutrophil Cytoplasmic Antibodies”[all fields] AND (“genes”[mesh] OR “genes”[all fields] OR “gene”[all fields] OR “genetic”[all fields] OR “genetics”[Subheading] OR “genetics”[all fields] OR “polymorphism, genetic”[mesh] OR “polymorphism”[all fields] OR “polymorphisms”[all fields] OR “dna”[mesh] OR “dna”[all fields] OR “genome”[mesh] OR “genome”[all fields] OR “genomes”[all fields] OR “genomics”[mesh] OR “genomics”[all fields] OR “Genetic Phenomena”[mesh] OR “Genetic Structures”[Mesh]))</p>

Supplementary table 1. Literature search strategies (Continued)

Database	Search Strategy
Embase	<p><i>Two strategies:</i></p> <p><u>1. AAV and genes</u> (ANCA associated vasculitis/ OR "ANCA-associated vasculitis".mp OR "ANCA associated vasculitis".mp OR "Anti-Neutrophil Cytoplasmic Antibody-Associated Vasculitis".mp OR "Pauci-Immune Vasculitis".mp OR "Pauci-Immune Vasculitis".mp OR "ANCA-Associated Vasculitides".mp OR "ANCA-Associated Vasculitide".mp OR "ANCA".mp AND vasculit*.mp) OR ("Anti-Neutrophil".mp AND "Cytoplasmic".mp AND "Antibody-Associated".mp AND vasculit*.mp) OR "Churg-Strauss Syndrome".mp OR "Churg Strauss Syndrome".mp OR "Allergic Granulomatous Angiitis".mp OR "Allergic Granulomatous Angiitides".mp OR "Allergic Angiitis".mp OR "Churg-Strauss Vasculitis".mp OR "eosinophilic granulomatosis with polyangiitis".mp OR "eosinophilic".mp AND "granulomatosis".mp OR "Wegener's Granulomatosis".mp OR "Microscopic Polyangiitis".mp OR "Wegener Granulomatosis".mp AND "polyangiitis".mp) OR "Wegener's Granulomatosis".mp OR "granulomatosis with polyangiitis".mp OR ("granulomatosis".mp AND "polyangiitis".mp) OR "anca".mp OR ("pr3".mp OR "mpo".mp) AND (exp Vasculitis/ OR vasculit*.mp OR "anca".mp)) OR neutrophil cytoplasmic antibody/ OR "Antineutrophil Cytoplasmic Antibody".mp OR "Anti-Neutrophil Cytoplasmic Antibody".mp OR "Anti Neutrophil Cytoplasmic Antibodies".mp AND (exp Gene / OR gene.mp OR genom*.mp OR genes.mp OR exp genetic polymorphism/ OR polymorphism*.mp OR dna.mp OR exp dna/ OR exp *Heridity/)</p>

Supplementary table 1. Literature search strategies (Continued)

Database	Search Strategy
Web of Science	<p><i>Two strategies:</i></p> <p><u>1. AAV and genes</u> TS=((“ANCA-associated vasculitis” OR “ANCA associated vasculitis” OR “Anti-Neutrophil Cytoplasmic Antibody-Associated Vasculitis” OR “Pauci-Immune Vasculitis” OR “Pauci Immune Vasculitis” OR “ANCA-Associated Vasculitides” OR “ANCA Associated Vasculitides” OR “ANCA-Associated Vasculite” OR “ANCA” AND vasculit*) OR (“Anti-Neutrophil” AND “Cytoplasmic” AND “Antibody-Associated” AND vasculit*) OR “Churg-Strauss Syndrome” OR “Allergic Angiitis” OR “eosinophilic granulomatosis with polyangiitis” OR “Allergic Granulomatous Angiitides” OR “Churg-Strauss Vasculitis” OR “Microscopic Polyangiitis” OR “Wegener Granulomatosis” OR “Wegener’s Granulomatosis” OR “granulomatosis with polyangiitis” OR “Microscopic Polyangiitis” OR “Microscopic Polyangiitis” OR “Microscopic Polyangiitis” OR “Wegener Granulomatosis” OR “Wegener’s Granulomatosis” OR “granulomatosis with polyangiitis” OR “neutrophil cytoplasmic antibody” OR “ANCA” OR (“pr-3” OR “mpo” AND (exp Vasculitis/ OR vasculit* OR “anca”)) OR “neutrophil cytoplasmic antibody” OR “Antineutrophil Cytoplasmic Antibody” OR “Anti-Neutrophil Cytoplasmic Antibody” OR “Anti Neutrophil Cytoplasmic Antibodies” OR “Antineutrophil Cytoplasmic Antibodies”) AND (gene OR genes OR genom* OR genetic* OR Polymorphism* OR dna))</p> <p><u>2. Vasculitis and genes (excluding AAV)</u> (TF=((vasculit*) AND (gene OR genes OR genom* OR genetic* OR Polymorphism* OR dna))) NOT (TS=((“ANCA-associated vasculitis” OR “ANCA associated vasculitis” OR “Anti-Neutrophil Cytoplasmic Antibody-Associated Vasculitis” OR “Pauci-Immune Vasculitis” OR “Pauci Immune Vasculitis” OR “ANCA associated vasculitis” OR “ANCA-Associated Vasculitides” OR “ANCA Associated Vasculitides” OR “ANCA-Associated Vasculite” OR “ANCA” AND vasculit*) OR (“Anti-Neutrophil” AND “Cytoplasmic” AND “Antibody-Associated” AND vasculit*) OR “Churg-Strauss Syndrome” OR “Allergic Angiitis” OR “eosinophilic granulomatous Angiitides” OR “Allergic Angiitis” OR “Churg-Strauss Vasculitis” OR “Microscopic Polyangiitis” OR “Wegener Granulomatosis” OR “Wegener’s Granulomatosis” OR “granulomatosis with polyangiitis” OR “Microscopic Polyangiitis” OR “Microscopic Polyangiitis” OR “Wegener Granulomatosis” OR “Wegener’s Granulomatosis” OR “granulomatosis with polyangiitis” OR “neutrophil cytoplasmic antibody” OR “ANCA” OR (“pr-3” OR “mpo” AND (exp Vasculitis/ OR vasculit* OR “anca”)) OR “neutrophil cytoplasmic antibody” OR “Antineutrophil Cytoplasmic Antibody” OR “Anti-Neutrophil Cytoplasmic Antibody” OR “Anti Neutrophil Cytoplasmic Antibodies” OR “Antineutrophil Cytoplasmic Antibodies”) AND (gene OR genes OR genom* OR genetic* OR Polymorphism* OR dna)))</p>

Supplementary table 2. Study characteristics of the included studies

Variants	Article	Study details			Definitions		Participants (n)			OR (95% CI)
		Design	Setting	Cases	Cases	Controls	Cases / Controls	Diagnoses	ANCA serotype	
CD226 rs763361 (T)	Wieczorek et al. 2009 ^{1*}	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy subjects	761/1226	GPA (642), EGPA (119)	PR3-ANCA (438), MPO-ANCA (32), ANCA negative (43)	1.24 (1.11 – 1.39)	
		Case-Control	UK	ACR and CHCC criteria	Geographically matched subjects from the general population	105/9337	GPA (all patients)	NR	1.17 (0.89 – 1.54)	
	Chung et al. 2012 ^{2**}	Case-Control	USA	ACR criteria	Geographically matched subjects with no (family) history of autoimmune disease	424/469	GPA (all patients)	PR3-ANCA (339), MPO-ANCA (44), ANCA negative (39)	1.08 (0.90 – 1.30)	
		Case-Control	Canada	ACR criteria	Geographically matched subjects with no history of autoimmune disease	456/1500	GPA (all patients)	NR	1.09 (0.94 – 1.26)	
CTLA-4 (AT)₈₆	Previously unpublished GWAS data ³ Lyons et al. 2012	Case-control (GWAS)	UK	EMA algorithm, supported by either serology or histology	Wellcome Trust Case Control Consortium (UK)	676/5366	GPA (394), MPA (156)	PR3-ANCA (326), MPO-ANCA (167)	1.08 (0.96 – 1.21)	
		Case-Control	Sweden	ACR criteria	Geographically and ethnically matched healthy subjects	32/109	GPA (all patients)	PR3-ANCA (29)	0.18 (0.10 – 0.35)	
	Tsuchiya et al. 2003 ⁵	Case-Control	Japan	CHCC and Japanese 1998 criteria	Geographically matched healthy subjects	49/111	MPA (all patients)	MPO-ANCA (all patients)	1.31 (0.76 – 2.25)	
	Zhou et al. 2004 ⁶	Case-Control	USA	ACR criteria	Geographically and ethnically matched healthy, unrelated subjects	117/123	GPA (all patients)	PR3-ANCA (82), MPO-ANCA (5), PR3- and MPO-ANCA positive (2), ANCA negative (25)	0.38 (0.26 – 0.55)	

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Article	Study details			Definitions		Participants (n)		OR (95% CI)
		Design	Setting	Cases	Controls	Cases / Controls	Diagnoses	ANCA serotype	
	Persson et al. 2013 ⁷	Case-Control	Sweden	ACR and CHCC criteria, EMA algorithm	Blood donors	105/200	GPA (61), MPA (44)	PR3-ANCA (62), MPO-ANCA (43)	0.72 (0.51 – 1.02)
<i>CTLA-4</i> (AT) ₁₀₂	Huang et al. 2000 ⁴	Case-Control	Sweden	ACR criteria	Geographically and ethnically matched healthy subjects	32/109	GPA (all patients)	PR3-ANCA (29)	1.71 (0.15 – 19.22)
	Tsuchiya et al. 2003 ⁵	Case-Control	Japan	CHCC and Japanese 1998 criteria	Geographically matched healthy subjects	49/111	MPA (all patients)	MPO-ANCA (all patients)	0.89 (0.47 – 1.68)
	Persson et al. 2013 ⁷	Case-Control	Sweden	ACR and CHCC criteria, EMA algorithm	Blood donors	105/200	GPA (61), MPA (44)	PR3-ANCA (62), MPO-ANCA (43)	1.73 (0.85 – 3.54)
	Huang et al. 2000 ⁴	Case-Control	Sweden	ACR criteria	Geographically and ethnically matched healthy subjects	32/109	GPA (all patients)	PR3-ANCA (29)	1.74 (0.96 – 3.13)
<i>CTLA-4</i> (AT) ₁₀₄	Tsuchiya et al. 2003 ⁵	Case-Control	Japan	CHCC and Japanese 1998 criteria	Geographically matched healthy subjects	49/111	MPA (all patients)	MPO-ANCA (all patients)	1.15 (0.70 – 1.91)
	Persson et al. 2013 ⁷	Case-Control	Sweden	ACR and CHCC criteria, EMA algorithm	Blood donors	105/200	GPA (61), MPA (44)	PR3-ANCA (62), MPO-ANCA (43)	1.09 (0.70 – 1.59)
	Huang et al. 2000 ⁴	Case-Control	Sweden	ACR criteria	Geographically and ethnically matched healthy subjects	32/109	GPA (all patients)	PR3-ANCA (29)	2.84 (0.74 – 10.91)
	Tsuchiya et al. 2003 ⁵	Case-Control	Japan	CHCC and Japanese 1998 criteria	Geographically matched healthy subjects	49/111	MPA (all patients)	MPO-ANCA (all patients)	0.41 (0.14 – 1.22)
<i>CTLA-4</i> (AT) ₁₀₆	Persson et al. 2013 ⁷	Case-Control	Sweden	ACR and CHCC criteria, EMA algorithm	Blood donors	105/200	GPA (61), MPA (44)	PR3-ANCA (62), MPO-ANCA (43)	2.63 (1.09 – 6.35)
	Huang et al. 2000 ⁴	Case-Control	Sweden	ACR criteria	Geographically and ethnically matched healthy subjects	32/109	GPA (all patients)	PR3-ANCA (29)	0.90 (0.42 – 1.95)

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Article	Study details		Definitions		Cases / Controls		Participants (n)		OR (95% CI)
		Design	Setting	Cases	Controls	Cases / Controls	Diagnoses	ANCA serotype	Allele level	
	Tsuchiya et al. 2003 ⁵	Case-Control	Japan	CHCC and Japanese 1998 criteria	Geographically matched healthy subjects	49/111	MPA (all patients)	MPO-ANCA (all patients)	0.89 (0.47 – 1.68)	
	Persson et al. 2013 ⁷	Case-Control	Sweden	ACR and CHCC criteria, EMA algorithm	Blood donors	105/200	GPA (61), MPA (44)	PR3-ANCA (62), MPO-ANCA (43)	1.78 (1.05 – 3.03)	
	Huang et al. 2000 ⁴	Case-Control	Sweden	ACR criteria	Geographically and ethnically matched healthy subjects	32/109	GPA (all patients)	PR3-ANCA (29)	0.48 (0.02-9.36)	
<i>CTLA-4</i> (AT) ₁₁₀	Tsuchiya et al. 2003 ⁵	Case-Control	Japan	CHCC and Japanese 1998 criteria	Geographically matched healthy subjects	49/111	MPA (all patients)	MPO-ANCA (all patients)	0.25 (0.01 – 4.62)	
	Persson et al. 2013 ⁷	Case-Control	Sweden	ACR and CHCC criteria, EMA algorithm	Blood donors	105/200	GPA (61), MPA (44)	PR3-ANCA (62), MPO-ANCA (43)	0.74 (0.34 – 1.61)	
	Huang et al. 2000 ⁴	Case-Control	Sweden	ACR criteria	Geographically and ethnically matched healthy subjects	32/109	GPA (all patients)	PR3-ANCA (29)	1.37 (0.26 – 7.26)	
<i>CTLA-4</i> (AT) ₁₁₆	Tsuchiya et al. 2003 ⁵	Case-Control	Japan	CHCC and Japanese 1998 criteria	Geographically matched healthy subjects	49/111	MPA (all patients)	MPO-ANCA (all patients)	0.75 (0.03 – 18.56)	
	Persson et al. 2013 ⁷	Case-Control	Sweden	ACR and CHCC criteria, EMA algorithm	Blood donors	105/200	GPA (61), MPA (44)	PR3-ANCA (62), MPO-ANCA (43)	0.540 (0.11 – 2.62)	
	Huang et al. 2000 ⁴	Case-Control	Sweden	ACR criteria	Geographically and ethnically matched healthy subjects	32/109	GPA (all patients)	PR3-ANCA (29)	3.44 (0.21 – 55.85)	
<i>CTLA-4</i> (AT) ₁₁₈	Tsuchiya et al. 2003 ⁵	Case-Control	Japan	CHCC and Japanese 1998 criteria	Geographically matched healthy subjects	49/111	MPA (all patients)	MPO-ANCA (all patients)	0.75 (0.08 – 7.33)	
	Persson et al. 2013 ⁷	Case-Control	Sweden	ACR and CHCC criteria, EMA algorithm	Blood donors	105/200	GPA (61), MPA (44)	PR3-ANCA (62), MPO-ANCA (43)	0.27 (0.06 – 1.18)	

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Article	Study details			Definitions		Participants (n)			OR (95% CI)
		Design	Setting	Cases	Controls	Cases / Controls	Diagnoses	ANCA serotype	Allele level	
<i>CTLA-4</i> (AT) ₁₂₂	Huang et al. 2000 ⁴	Case-Control	Sweden	ACR criteria	Geographically and ethnically matched healthy subjects	32/109	GPA (all patients)	PR3-ANCA (29)	10.32 (0.42 – 256.50)	
	Tsuchiya et al. 2003 ⁵	Case-Control	Japan	CHCC and Japanese 1998 criteria	Geographically matched healthy subjects	49/111	MPA (all patients)	MPO-ANCA (all patients)	2.32 (0.57 – 9.47)	
	Persson et al. 2013 ⁷	Case-Control	Sweden	ACR and CHCC criteria, EMA algorithm	Blood donors	105/200	GPA (61), MPA (44)	PR3-ANCA (62), MPO-ANCA (43)	0.47 (0.05 – 4.27)	
<i>CTLA-4</i> (AT) ₁₂₄	Huang et al. 2000 ⁴	Case-Control	Sweden	ACR criteria	Geographically and ethnically matched healthy subjects	32/109	GPA (all patients)	PR3-ANCA (29)	14.47 (1.59 – 131.86)	
	Tsuchiya et al. 2003 ⁵	Case-Control	Japan	CHCC and Japanese 1998 criteria	Geographically matched healthy subjects	49/111	MPA (all patients)	MPO-ANCA (all patients)	2.28 (0.14 – 36.59)	
	Persson et al. 2013 ⁷	Case-Control	Sweden	ACR and CHCC criteria, EMA algorithm	Blood donors	105/200	GPA (61), MPA (44)	PR3-ANCA (62), MPO-ANCA (43)	0.63 (0.13 – 3.16)	
<i>CTLA-4</i> (AT) ₁₂₆	Huang et al. 2000 ⁴	Case-Control	Sweden	ACR criteria	Geographically and ethnically matched healthy subjects	32/109	GPA (all patients)	PR3-ANCA (29)	4.78 (1.04 – 21.93)	
	Tsuchiya et al. 2003 ⁵	Case-Control	Japan	CHCC and Japanese 1998 criteria	Geographically matched healthy subjects	49/111	MPA (all patients)	MPO-ANCA (all patients)	0.45 (0.05 – 3.88)	
	Persson et al. 2013 ⁷	Case-Control	Sweden	ACR and CHCC criteria, EMA algorithm	Blood donors	105/200	GPA (61), MPA (44)	PR3-ANCA (62), MPO-ANCA (43)	0.81 (0.21 – 3.18)	
<i>CTLA-4</i> (AT) ₁₂₈	Huang et al. 2000 ⁴	Case-Control	Sweden	ACR criteria	Geographically and ethnically matched healthy subjects	32/109	GPA (all patients)	PR3-ANCA (29)	1.12 (0.05 – 27.93)	
	Tsuchiya et al. 2003 ⁵	Case-Control	Japan	CHCC and Japanese 1998 criteria	Geographically matched healthy subjects	49/111	MPA (all patients)	MPO-ANCA (all patients)	6.98 (0.72 – 67.95)	

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Article		Study details		Definitions		Cases / Controls	Participants (n)		OR (95% CI)
	Design	Setting	Cases	Controls	Diagnoses	ANCA serotype		Allele level		
CTLA-4 rs231775 (G)	Persson et al. 2013 ⁷	Sweden	ACR and CHCC criteria, EMA algorithm	Blood donors	105/200	GPA (61), MPA (44)	PR3-ANCA (62), MPO-ANCA (43)	0.69 (0.22 – 2.18)		
	Slot et al. 2008 ⁸	The Netherlands	CHCC criteria	Geographically and ethnically matched healthy subjects	104/185	GPA (50), MPA (24), EGPA (7), RLV (21)	PR3-ANCA (49), MPO-ANCA (34), PR3- and MPO-ANCA positive (2), ANCA negative (17)	1.53 (1.09 – 2.17)		
	Kamesh et al. 2009 ⁹	UK	CHCC criteria	Ethnically matched healthy subjects with no (family) history of autoimmune disease	222/629	GPA (116), MPA (96)	NR	1.32 (1.06 – 1.65)		
	Previously unpublished Lyons et al. 2012 GWAS data ³	UK	EMA algorithm, supported by either serology or histology	Wellcome Trust Case Control Consortium (UK)	676/5365	GPA (394), MPA (156)	PR3-ANCA (326), MPO-ANCA (167)	1.09 (0.97 – 1.22)		
CTLA-4 rs3087243 (A)	Kamesh et al. 2009 ⁹	UK	CHCC criteria	Ethnically matched healthy subjects with no (family) history of autoimmune disease	222/629	GPA (116), MPA (96)	NR	0.67 (0.54 – 0.83)		
	Chung et al. 2012 ^{2*}	USA	ACR criteria	Geographically matched subjects with no (family) history of autoimmune disease	424/469	GPA (all patients)	PR3-ANCA (339), MPO-ANCA (44), ANCA negative (39)	0.80 (0.66 – 0.97)		
	Case-Control	Canada	ACR criteria	Geographically matched subjects with no history of autoimmune disease	456/1500	GPA (all patients)	NR	0.78 (0.67 – 0.91)		

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Article	Study details			Definitions		Participants (n)			OR (95% CI)
		Design	Setting	Cases	Controls	Cases / Controls	Diagnoses	ANCA serotype	Allele level	
FCAR rs16986050 (G)	Previously unpublished Lyons et al. 2012 GWAS data ³	Case-control (GWAS)	UK	EMA algorithm, supported by either serology or histology	Wellcome Trust Case Control Consortium (UK)	913/5257	GPA (565), MPA (262)	PR3-ANCA (478), MPO-ANCA (264)	0.85 (0.77 - 0.94)	
	Kelley et al. 2011 ^{10**}	Case-Control	USA	ACR and CHCC criteria	Geographically matched subjects with no (family) history of autoimmune disease	445/413	GPA (all patients)	NR	0.65 (0.50 – 0.83)	
FCGR2A rs1801274 (C)	Previously unpublished Lyons et al. 2012 GWAS data ³	Case-control (GWAS)	UK	EMA algorithm, supported by either serology or histology	Wellcome Trust Case Control Consortium (UK)	676/5365	GPA (394), MPA (156)	PR3-ANCA (326), MPO-ANCA (167)	1.07 (0.93 – 1.24)	
	Edberg et al. 1997 ¹¹	Case-Control	USA, Chile, Germany, Canada	ACR and CHCC criteria	Ethnically matched healthy volunteers	147/149	GPA (all patients)	NR	0.85 (0.61 – 1.17)	
(C)	Dijstelbloem et al. 1999 ¹²	Case-Control	The Netherlands	ACR criteria	Ethnically matched healthy blood donors	91/154	GPA (all patients)	PR3-ANCA (all patients)	0.98 (0.68 – 1.41)	
	Tse et al. 1999 ¹³	Case-Control	UK	CHCC criteria	Ethnically matched healthy subjects	107/100	GPA (48), MPA (54), EGPA (1), PAN (4)	PR3-ANCA (75), MPO-ANCA (32)	1.00 (0.68 – 1.48)	
	Tsuchiya et al. 2003 ⁵	Case-Control	Japan	CHCC and Japanese 1998 criteria	Geographically matched healthy subjects	50/303	MPA (all patients)	MPO-ANCA (all patients)	0.68 (0.37 – 1.24)	

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Article		Study details		Definitions		Cases / Controls		Participants (n)		OR (95% CI)	
	Design	Setting	Cases	Controls	Cases / Controls	Diagnoses	ANCA serotype	Allele level				
FCGR3A rs396991 (G)	Previously unpublished Lyons et al. 2012 GWAS data ³	UK	EMA algorithm, supported by either serology or histology	Wellcome Trust Case Control Consortium (UK)	676/5365	GPA (394), MPA (156)	PR3-ANCA (326), MPO-ANCA (167)	0.89 (0.79 – 0.99)				
	Persson et al. 2013 ⁷	Sweden	ACR and CHCC criteria, EMA algorithm	Blood donors	105/200	GPA (61), MPA (44)	PR3-ANCA (62), MPO-ANCA (43)	1.03 (0.73 – 1.43)				
FCGR3B (NA1)	Dijstelbloem et al. 1999 ¹²	The Netherlands	ACR criteria	Ethnically matched healthy blood donors	91/154	GPA (all patients)	PR3-ANCA (all patients)	1.30 (0.89 – 1.91)				
	Tsuchiya et al. 2003 ⁵	Japan	CHCC and Japanese 1998 criteria	Geographically matched healthy subjects	50/303	MPA (all patients)	MPO-ANCA (all patients)	0.65 (0.39 – 1.07)				
FCGR3B (NA1)	Dijstelbloem et al. 1999 ¹²	The Netherlands	ACR criteria	Ethnically matched healthy blood donors	91/154	GPA (all patients)	PR3-ANCA (all patients)	1.05 (0.72 – 1.53)				
	Tse et al. 2000 ⁴	UK	CHCC criteria	Ethnically matched healthy subjects	101/100	GPA (45), MPA (52), EGPA (1), PAN (3)	PR3-ANCA (61), MPO-ANCA (30)	1.24 (0.8 – 1.87)				
GHSR rs509035 (A)	Tsuchiya et al. 2003 ⁵	Japan	CHCC and Japanese 1998 criteria	Geographically matched healthy subjects	50/303	MPA (all patients)	MPO-ANCA (all patients)	1.29 (0.82 – 2.03)				
	Kelley et al. 2011 ¹⁰	USA	ACR and CHCC criteria	Geographically matched subjects with no (family) history of autoimmune disease	673/413	GPA (all patients)	NR	0.86 (0.72 – 1.03)				
GHSR rs509035 (A)	Wieczorek et al. 2010 ^{15**}	Germany	CHCC criteria	Geographically matched healthy subjects	454/820	GPA (all patients)	NR	1.22 (1.02 – 1.45)				
		Germany	CHCC criteria	Geographically matched healthy subjects	179/516	GPA (all patients)	NR	0.94 (0.72 – 1.23)				

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Article	Study details			Definitions		Controls		Participants (n)		OR (95% CI)
		Design	Setting	Cases	Controls	Cases /	Diagnoses	ANCA serotype	Allele level		
	Previously unpublished Lyons et al. 2012 GWAS data ³	Case-control (GWAS)	UK	EMA algorithm, supported by either serology or histology	Wellcome Trust Case Control Consortium (UK)	914/5259	GPA (565), MPA (262)	PR3-ANCA (478), MPO-ANCA (264)	1.00 (0.90 – 1.11)		
GHSR	Wieczorek et al. 2010 ^{15**}	Case-Control	Germany	CHCC criteria	Geographically matched healthy subjects	449/813	GPA (all patients)	NR	1.17 (0.98 – 1.40)		
rs519384 (A)		Case-Control	Germany	CHCC criteria	Geographically matched healthy subjects	179/515	GPA (all patients)	NR	1.01 (0.77 – 1.33)		
GHSR	Wieczorek et al. 2010 ^{15**}	Case-Control	Germany	CHCC criteria	Geographically matched healthy subjects	442/812	GPA (all patients)	NR	1.21 (1.01 – 1.44)		
rs572169 (A)		Case-Control	Germany	CHCC criteria	Geographically matched healthy subjects	178/492	GPA (all patients)	NR	0.96 (0.73 – 1.25)		
HLA-A1	Strimlan et al. 1978 ¹⁶	Case-Control	USA	Histologically proven AAV	Ethnically matched healthy blood donors	31/701	GPA (all patients)	NR	1.53 (0.81 – 2.87)		
	Murty et al. 1991 ¹⁷	Case-Control	Ireland, UK	Histologically proven AAV	Geographically matched healthy subjects	41/700	GPA (all patients)	NR	0.84 (0.47 – 1.52)		
	Von Vietinghoff et al. 2006 ¹⁸	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	51/51	GPA (all patients)	NR	0.92 (0.42 – 2.03)		
	Stassen et al. 2009 ¹⁹	Case-Control	The Netherlands	CHCC criteria	Geographically matched healthy blood donors	304/5872	GPA (241), MPA (30), EGPA (12), RVL (21)	PR3-ANCA (218), MPO-ANCA (64), other (4), ANCA negative (18)	1.08 (0.87 – 1.35)		
HLA-A2	Strimlan et al. 1978 ¹⁶	Case-Control	USA	Histologically proven AAV	Ethnically matched healthy blood donors	31/701	GPA (all patients)	NR	0.78 (0.42 – 1.45)		
	Murty et al. 1991 ¹⁷	Case-Control	Ireland, UK	Histologically proven AAV	Geographically matched healthy subjects	41/700	GPA (all patients)	NR	1.04 (0.62 – 1.73)		

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Article		Study details		Definitions		Cases / Controls		Participants (n)		OR (95% CI)	
	Article	Design	Setting	Cases	Controls	Cases / Controls	Diagnoses	ANCA serotype	Allele level			
	Nakamaru et al. 1996 ²⁰	Case-Control	Japan	Clinical, histological, serological criteria	Geographically matched healthy subjects	16/472	GPA (all patients)	c-ANCA (all patients)	0.90 (0.37 – 2.23)			
	Von Vietinghoff et al. 2006 ¹⁸	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	51/51	GPA (all patients)	NR	0.87 (0.48 – 1.58)			
	Stassen et al. 2009 ¹⁹	Case-Control	The Netherlands	CHCC criteria	Geographically matched healthy blood donors	304/5872	GPA (241), MPA (30), EGPA (12), RVL (21)	PR3-ANCA (218), MPO-ANCA (6+), other (4), ANCA negative (18)	0.98 (0.81 – 1.17)			
HLA-A3	Strimlan et al. 1978 ¹⁶	Case-Control	USA	Histologically proven AAV	Ethnically matched healthy blood donors	31/701	GPA (all patients)	NR	1.73 (0.90 – 3.31)			
	Murty et al. 1991 ¹⁷	Case-Control	Ireland, UK	Histologically proven AAV	Geographically matched healthy subjects	41/700	GPA (all patients)	NR	1.51 (0.86 – 2.66)			
	Von Vietinghoff et al. 2006 ¹⁸	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	51/51	GPA (all patients)	NR	1.88 (0.89 – 3.98)			
	Stassen et al. 2009 ¹⁹	Case-Control	The Netherlands	CHCC criteria	Geographically matched healthy blood donors	304/5872	GPA (241), MPA (30), EGPA (12), RVL (21)	PR3-ANCA (218), MPO-ANCA (6+), other (4), ANCA negative (18)	1.04 (0.84 – 1.31)			
	Strimlan et al. 1978 ¹⁶	Case-Control	USA	Histologically proven AAV	Ethnically matched healthy blood donors	31/701	GPA (all patients)	NR	1.09 (0.46 – 2.59)			
HLA-A9	Katz et al. 1979 ²¹	Case-Control	USA	Clinical and histological criteria	Ethnically matched subjects	31/418	GPA (all patients)	NR	69.13 (3.28 – 456.19)			

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Article	Study details			Definitions		Participants (n)			OR (95% CI)
		Design	Setting	Cases	Controls	Cases / Controls	Diagnoses	ANCA serotype	Allele level	
	Stassen et al. 2009 ¹⁹	Case-Control	The Netherlands	CHCC criteria	Geographically matched healthy blood donors	304/5872	GPA (241), MPA (30), EGPA (12), RVL (21)	PR3-ANCA (218), MPO-ANCA (64), other (4), ANCA negative (18)	0.79 (0.58 – 1.06)	
HLA-A10	Strimlan et al. 1978 ¹⁶	Case-Control	USA	Histologically proven AAV	Ethnically matched healthy blood donors	31/701	GPA (all patients)	NR	1.06 (0.32 – 3.48)	
	Katz et al. 1979 ²¹	Case-Control	USA	Clinical and histological criteria	Ethnically matched subjects	31/418	GPA (all patients)	NR	69.13 (3.28 – 1456.19)	
	Stassen et al. 2009 ¹⁹	Case-Control	The Netherlands	CHCC criteria	Geographically matched healthy blood donors	304/5872	GPA (241), MPA (30), EGPA (12), RVL (21)	PR3-ANCA (218), MPO-ANCA (64), other (4), ANCA negative (18)	0.61 (0.33 – 1.12)	
HLA-A11	Strimlan et al. 1978 ¹⁶	Case-Control	USA	Histologically proven AAV	Ethnically matched healthy blood donors	31/607	GPA (all patients)	NR	1.96 (0.81 – 4.72)	
	Murty et al. 1991 ¹⁷	Case-Control	Ireland, UK	Histologically proven AAV	Geographically matched healthy subjects	41/700	GPA (all patients)	NR	0.68 (0.24 – 1.90)	
	Nakamaru et al. 1996 ²⁰	Case-Control	Japan	Clinical, histological, serological criteria	Geographically matched healthy subjects	16/472	GPA (all patients)	c-ANCA (all patients)	0.71 (0.17 – 3.03)	
	Papasteriades et al. 1997 ²²	Case-Control	Greece	Histologically proven AAV	Geographically and ethnically matched healthy subjects	23/405	MPA (all patients)	NR	2.97 (1.19 – 7.41)	
	Von Vietinghoff et al. 2006 ¹⁸	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	51/51	GPA (all patients)	NR	0.43 (0.14 – 1.27)	

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Article	Study details		Definitions		Participants (n)		OR (95% CI)	
		Design	Setting	Cases	Controls	Cases / Controls	Diagnoses	ANCA serotype	Allele level
	Stassen et al. 2009 ¹⁹	Case-Control	The Netherlands	CHCC criteria	Geographically matched healthy blood donors	304/5872	GPA (241), MPA (30), EGPA (12), RVL (21)	PR3-ANCA (218), MPO-ANCA (64), other (+), ANCA negative (18)	0.62 (0.39 – 0.99)
HLA-A24	Murty et al. 1991 ¹⁷	Case-Control	Ireland, UK	Histologically proven AAV	Geographically matched healthy subjects	41/700	GPA (all patients)	NR	0.91 (0.36 – 2.31)
	Nakamaru et al. 1996 ²⁰	Case-Control	Japan	Clinical, histological, serological criteria	Geographically matched healthy subjects	16/472	GPA (all patients)	c-ANCA (all patients)	0.75 (0.34 – 1.65)
	Von Vietinghoff et al. 2006 ¹⁸	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	51/51	GPA (all patients)	NR	1.81 (0.51 – 6.37)
HLA-A25	Murty et al. 1991 ¹⁷	Case-Control	Ireland, UK	Histologically proven AAV	Geographically matched healthy subjects	41/700	GPA (all patients)	NR	1.43 (0.33 – 6.17)
	Von Vietinghoff et al. 2006 ¹⁸	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	51/51	GPA (all patients)	NR	2.09 (0.61 – 7.16)
HLA-A26	Murty et al. 1991 ¹⁷	Case-Control	Ireland, UK	Histologically proven AAV	Geographically matched healthy subjects	41/700	GPA (all patients)	NR	0.68 (0.09 – 5.07)
	Nakamaru et al. 1996 ²⁰	Case-Control	Japan	Clinical, histological, serological criteria	Geographically matched healthy subjects	16/472	GPA (all patients)	c-ANCA (all patients)	2.36 (1.00. – 5.60)
	Papasteriades et al. 1997 ²²	Case-Control	Greece	Histologically proven AAV	Geographically and ethnically matched healthy subjects	23/405	MPA (all patients)	NR	2.49 (1.01 – 6.18)
	Von Vietinghoff et al. 2006 ¹⁸	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	51/51	GPA (all patients)	NR	0.39 (0.07 – 2.05)
HLA-A28	Sririmlan et al. 1978 ¹⁶	Case-Control	USA	Histologically proven AAV	Ethnically matched healthy blood donors	31/701	GPA (all patients)	NR	0.40 (0.02 – 6.68)

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Article	Study details			Definitions		Participants (n)			OR (95% CI) Allele level
		Design	Setting	Cases	Controls	Cases / Controls	Diagnoses	ANCA serotype		
	Murty et al. 1991 ¹⁷	Case-Control	Ireland, UK	Histologically proven AAV	Geographically matched healthy subjects	41/700	GPA (all patients)	NR	0.32 (0.04 – 2.34)	
	Stassen et al. 2009 ¹⁹	Case-Control	The Netherlands	CHCC criteria	Geographically matched healthy blood donors	304/5872	GPA (2+), MPA (30), EGPA (12), RVL (21)	PR3-ANCA (218), MPO-ANCA (6+), other (+), ANCA negative (18)	0.88 (0.57 – 1.38)	
HLA-A29	Strimlan et al. 1978 ¹⁶	Case-Control	USA	Histologically proven AAV	Ethnically matched healthy blood donors	31/701	GPA (all patients)	NR	2.11 (0.68 – 6.57)	
	Murty et al. 1991 ¹⁷	Case-Control	Ireland, UK	Histologically proven AAV	Geographically matched healthy subjects	41/700	GPA (all patients)	NR	0.31 (0.04 – 2.25)	
	Von Vietinghoff et al. 2006 ¹⁸	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	51/51	GPA (all patients)	NR	0.66 (0.11 – 4.04)	
HLA-A30	Murty et al. 1991 ¹⁷	Case-Control	Ireland, UK	Histologically proven AAV	Geographically matched healthy subjects	41/700	GPA (all patients)	NR	0.68 (0.09 – 5.07)	
	Von Vietinghoff et al. 2006 ¹⁸	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	51/51	GPA (all patients)	NR	2.02 (0.18 – 22.63)	
HLA-A31	Murty et al. 1991 ¹⁷	Case-Control	Ireland, UK	Histologically proven AAV	Geographically matched healthy subjects	41/700	GPA (all patients)	NR	1.07 (0.25 – 4.54)	
	Nakamaru et al. 1996 ²⁰	Case-Control	Japan	Clinical, histological, serological criteria	Geographically matched healthy subjects	16/472	GPA (all patients)	c-ANCA (all patients)	1.55 (0.46 – 5.24)	
	Von Vietinghoff et al. 2006 ¹⁸	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	51/51	GPA (all patients)	NR	0.33 (0.03 – 3.20)	
HLA-A32	Murty et al. 1991 ¹⁷	Case-Control	Ireland, UK	Histologically proven AAV	Geographically matched healthy subjects	41/700	GPA (all patients)	NR	3.73 (1.24 – 11.22)	

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Article		Study details		Definitions		Cases / Controls		Participants (n)		OR (95% CI)	
	Design	Setting	Cases	Controls	Cases / Controls	Diagnoses	ANCA serotype	Allele level				
HLA-B5	Nakamaru et al. 1996 ²⁰	Japan	Clinical, histological, serological criteria	Geographically matched healthy subjects	16/472	GPA (all patients)	c-ANCA (all patients)	2.07 (0.60 – 7.04)				
	Von Vietinghoff et al. 2006 ¹⁸	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	51/51	GPA (all patients)	NR	0.24 (0.03 – 2.21)				
	Strimlan et al. 1978 ¹⁶	USA	Histologically proven AAV	Ethnically matched healthy blood donors	31/701	GPA (all patients)	NR	0.93 (0.28 – 3.02)				
	Stassen et al. 2009 ¹⁹	The Netherlands	CHCC criteria	Geographically matched healthy blood donors	304/5872	GPA (2+), MPA (30), EGPA (12), RVL (21)	PR3-ANCA (218), MPO-ANCA (6+), other (+), ANCA negative (18)	0.56 (0.35 – 0.89)				
	Strimlan et al. 1978 ¹⁶	USA	Histologically proven AAV	Ethnically matched healthy blood donors	31/701	GPA (all patients)	NR	1.54 (0.77 – 3.08)				
HLA-B7	Murty et al. 1991 ¹⁷	Ireland, UK	Histologically proven AAV	Geographically matched healthy subjects	41/700	GPA (all patients)	NR	1.74 (1.02 – 2.96)				
	Von Vietinghoff et al. 2006 ¹⁸	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	51/51	GPA (all patients)	NR	1.07 (0.52 – 2.22)				
	Stassen et al. 2009 ¹⁹	The Netherlands	CHCC criteria	Geographically matched healthy blood donors	304/5872	GPA (2+), MPA (30), EGPA (12), RVL (21)	PR3-ANCA (218), MPO-ANCA (6+), other (+), ANCA negative (18)	1.00 (0.80 – 1.26)				
	Strimlan et al. 1978 ¹⁶	USA	Histologically proven AAV	Ethnically matched healthy blood donors	31/701	GPA (all patients)	NR	1.24 (0.60 – 2.56)				
	Katz et al. 1979 ²¹	USA	Clinical and histological criteria	Ethnically matched subjects	31/418	GPA (all patients)	NR	2.30 (1.18 – 4.50)				

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Article	Study details			Definitions		Cases / Controls		Participants (n)		OR (95% CI)
		Design	Setting	Cases	Controls	Cases / Controls	Diagnoses	ANCA serotype	Allele level		
	Elkon et al. 1983 ²³	Case-Control	UK	Clinical and histological criteria	Geographically matched subjects	17/113	GPA (all patients)	NR	2.47 (1.01 – 6.05)		
	Murty et al. 1991 ¹⁷	Case-Control	Ireland, UK	Histologically proven AAV	Geographically matched healthy subjects	41/700	GPA (all patients)	NR	1.03 (0.55 – 1.93)		
	Von Vietinghoff et al. 2006 ¹⁸	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	51/51	GPA (all patients)	NR	3.27 (1.02 – 10.50)		
	Stassen et al. 2009 ¹⁹	Case-Control	The Netherlands	CHCC criteria	Geographically matched healthy blood donors	304/5872	GPA (241), MPA (30), EGPA (12), RVL (21)	PR3-ANCA (218), MPO-ANCA (64), other (+), ANCA negative (18)	1.11 (0.88 – 1.40)		
	Strimlan et al. 1978 ¹⁶	Case-Control	USA	Histologically proven AAV	Ethnically matched healthy blood donors	31/701	GPA (all patients)	NR	1.01 (0.47 – 2.15)		
	Stassen et al. 2009 ¹⁹	Case-Control	The Netherlands	CHCC criteria	Geographically matched healthy blood donors	304/5872	GPA (241), MPA (30), EGPA (12), RVL (21)	PR3-ANCA (218), MPO-ANCA (64), other (+), ANCA negative (18)	0.96 (0.74 – 1.24)		
HLA-B13	Strimlan et al. 1978 ¹⁶	Case-Control	USA	Histologically proven AAV	Ethnically matched healthy blood donors	31/701	GPA (all patients)	NR	0.87 (0.12 – 6.50)		
	Murty et al. 1991 ¹⁷	Case-Control	Ireland, UK	Histologically proven AAV	Geographically matched healthy subjects	41/700	GPA (all patients)	NR	2.09 (0.62 – 7.07)		
	Von Vietinghoff et al. 2006 ¹⁸	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	51/51	GPA (all patients)	NR	0.79 (0.21 – 3.03)		
	Stassen et al. 2009 ¹⁹	Case-Control	The Netherlands	CHCC criteria	Geographically matched healthy blood donors	304/5872	GPA (241), MPA (30), EGPA (12), RVL (21)	PR3-ANCA (218), MPO-ANCA (64), other (+), ANCA negative (18)	0.63 (0.31 – 1.28)		

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Article	Study details		Definitions		Cases / Controls		Participants (n)		OR (95% CI)
		Design	Setting	Cases	Controls	Cases / Controls	Diagnoses	ANCA serotype	Allele level	
HLA-B14	Strimlan et al. 1978 ¹⁶	Case-Control	USA	Histologically proven AAV	Ethnically matched healthy blood donors	31/701	GPA (all patients)	NR	0.32 (0.02 – 5.24)	
	Murty et al. 1991 ¹⁷	Case-Control	Ireland, UK	Histologically proven AAV	Geographically matched healthy subjects	41/700	GPA (all patients)	NR	0.51 (0.16 – 1.63)	
	Von Vietinghoff et al. 2006 ¹⁸	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	51/51	GPA (all patients)	NR	1.52 (0.25 – 9.26)	
	Stassen et al. 2009 ¹⁹	Case-Control	The Netherlands	CHCC criteria	Geographically matched healthy blood donors	304/5872	GPA (241), MPA (30), EGPA (12), RVL (21)	PR3-ANCA (218), MPO-ANCA (64), other (4), ANCA negative (18)	1.16 (0.63 – 2.14)	
HLA-B15	Von Vietinghoff et al. 2006 ¹⁸	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	51/51	GPA (all patients)	NR	1.38 (0.55 – 3.43)	
	Stassen et al. 2009 ¹⁹	Case-Control	The Netherlands	CHCC criteria	Geographically matched healthy blood donors	304/5872	GPA (241), MPA (30), EGPA (12), RVL (21)	PR3-ANCA (218), MPO-ANCA (64), other (4), ANCA negative (18)	0.98 (0.74 – 1.29)	
	Strimlan et al. 1978 ¹⁶	Case-Control	USA	Histologically proven AAV	Ethnically matched healthy blood donors	31/549	GPA (all patients)	NR	1.16 (0.35 – 3.85)	
HLA-B18	Murty et al. 1991 ¹⁷	Case-Control	Ireland, UK	Histologically proven AAV	Geographically matched healthy subjects	41/700	GPA (all patients)	NR	0.53 (0.13 – 2.21)	
	Von Vietinghoff et al. 2006 ¹⁸	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	51/51	GPA (all patients)	NR	0.78 (0.296 – 2.072)	
	Stassen et al. 2009 ¹⁹	Case-Control	The Netherlands	CHCC criteria	Geographically matched healthy blood donors	304/5872	GPA (241), MPA (30), EGPA (12), RVL (21)	PR3-ANCA (218), MPO-ANCA (64), other (4), ANCA negative (18)	0.75 (0.435 – 1.283)	

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Article	Study details			Definitions		Participants (n)			OR (95% CI)
		Design	Setting	Cases	Controls	Cases / Controls	Diagnoses	ANCA serotype	Allele level	
HLA-B27	Sirimlan et al. 1978 ¹⁶	Case-Control	USA	Histologically proven AAV	Ethnically matched healthy blood donors	31/600	GPA (all patients)	NR	0.74 (0.18 – 3.093)	
	Murty et al. 1991 ¹⁷	Case-Control	Ireland, UK	Histologically proven AAV	Geographically matched healthy subjects	41/700	GPA (all patients)	NR	0.60 (0.14 – 2.503)	
	Von Vietinghoff et al. 2006 ¹⁸	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	51/51	GPA (all patients)	NR	0.06 (0.00 – 1.102)	
	Stassen et al. 2009 ¹⁹	Case-Control	The Netherlands	CHCC criteria	Geographically matched healthy blood donors	304/5872	GPA (241), MPA (30), EGPA (12), RVL (21)	PR3-ANCA (218), MPO-ANCA (64), other (4), ANCA negative (18)	0.77 (0.48 – 1.247)	
HLA-B35	Murty et al. 1991 ¹⁷	Case-Control	Ireland, UK	Histologically proven AAV	Geographically matched healthy subjects	41/700	GPA (all patients)	NR	0.45 (0.11 – 1.86)	
	Nakamaru et al. 1996 ²⁰	Case-Control	Japan	Clinical, histological, serological criteria	Geographically matched healthy subjects	16/472	GPA (all patients)	c-ANCA (all patients)	0.80 (0.187 – 3.40)	
	Von Vietinghoff et al. 2006 ¹⁸	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	51/51	GPA (all patients)	NR	0.85 (0.28 – 2.62)	
	Stassen et al. 2009 ¹⁹	Case-Control	The Netherlands	CHCC criteria	Geographically matched healthy blood donors	304/5872	GPA (241), MPA (30), EGPA (12), RVL (21)	PR3-ANCA (218), MPO-ANCA (64), other (4), ANCA negative (18)	0.81 (0.59 – 1.12)	
HLA-B37	Von Vietinghoff et al. 2006 ¹⁸	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	51/51	GPA (all patients)	NR	0.14 (0.01 – 2.72)	

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Article	Study details		Definitions		Cases / Controls		Participants (n)		OR (95% CI)	
		Design	Setting	Cases	Controls	Cases / Controls	Diagnoses	ANCA serotype	Allele level		
HLA-B39	Stassen et al. 2009 ¹⁹	Case-Control	The Netherlands	CHCC criteria	Geographically matched healthy blood donors	304/5872	GPA (241), MPA (30), EGPA (12), RVL (21)	PR3-ANCA (218), MPO-ANCA (64), other (+), ANCA negative (18)	1.07 (0.58 – 1.98)		
	Nakamaru et al. 1996 ²⁰	Case-Control	Japan	Clinical, histological, serological criteria	Geographically matched healthy subjects	16/472	GPA (all patients)	c-ANCA (all patients)	0.86 (0.11 – 6.51)		
	Von Vietinghoff et al. 2006 ¹⁸	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	51/51	GPA (all patients)	NR	0.33 (0.01 – 8.20)		
HLA-B40	Von Vietinghoff et al. 2006 ¹⁸	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	51/51	GPA (all patients)	NR	1.65 (0.52 – 5.23)		
	Stassen et al. 2009 ¹⁹	Case-Control	The Netherlands	CHCC criteria	Geographically matched healthy blood donors	304/5872	GPA (241), MPA (30), EGPA (12), RVL (21)	PR3-ANCA (218), MPO-ANCA (64), other (+), ANCA negative (18)	0.90 (0.67 – 1.22)		
	Murty et al. 1991 ¹⁷	Case-Control	Ireland, UK	Histologically proven AAV	Geographically matched healthy subjects	41/700	GPA (all patients)	NR	1.07 (0.58 – 2.00)		
HLA-B49	Nakamaru et al. 1996 ²⁰	Case-Control	Japan	Clinical, histological, serological criteria	Geographically matched healthy subjects	16/472	GPA (all patients)	c-ANCA (all patients)	2.50 (0.845 – 7.403)		
	Von Vietinghoff et al. 2006 ¹⁸	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	51/51	GPA (all patients)	NR	3.27 (1.02 – 10.50)		
	Murty et al. 1991 ¹⁷	Case-Control	Ireland, UK	Histologically proven AAV	Geographically matched healthy subjects	41/700	GPA (all patients)	NR	1.27 (0.30 – 5.44)		
	Von Vietinghoff et al. 2006 ¹⁸	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	51/51	GPA (all patients)	NR	2.02 (0.18 – 22.63)		

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Article	Study details			Definitions		Participants (n)			OR (95% CI)
		Design	Setting	Cases	Controls	Cases / Controls	Diagnoses	ANCA serotype	Allele level	
HLA-B51	Nakamaru et al. 1996 ²⁰	Case-Control	Japan	Clinical, histological, serological criteria	Geographically matched healthy subjects	16/472	GPA (all patients)	c-ANCA (all patients)	1.87 (0.64 – 5.49)	
	Von Vietinghoff et al. 2006 ¹⁸	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	51/51	GPA (all patients)	NR	0.61 (0.19 – 1.92)	
HLA-B55	Nakamaru et al. 1996 ²⁰	Case-Control	Japan	Clinical, histological, serological criteria	Geographically matched healthy subjects	16/472	GPA (all patients)	c-ANCA (all patients)	7.35 (2.33 – 23.14)	
	Von Vietinghoff et al. 2006 ¹⁸	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	51/51	GPA (all patients)	NR	0.07 (0.00 – 1.30)	
HLA-B57	Murty et al. 1991 ¹⁷	Case-Control	Ireland, UK	Histologically proven AAV	Geographically matched healthy subjects	41/700	GPA (all patients)	NR	2.78 (1.05 – 7.32)	
	Von Vietinghoff et al. 2006 ¹⁸	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	51/51	GPA (all patients)	NR	1.00 (0.06 – 16.21)	
HLA-B60	Murty et al. 1991 ¹⁷	Case-Control	Ireland, UK	Histologically proven AAV	Geographically matched healthy subjects	41/700	GPA (all patients)	NR	3.51 (0.99 – 12.36)	
	Nakamaru et al. 1996 ²⁰	Case-Control	Japan	Clinical, histological, serological criteria	Geographically matched healthy subjects	16/472	GPA (all patients)	c-ANCA (all patients)	1.19 (0.28 – 5.13)	
HLA-B62	Murty et al. 1991 ¹⁷	Case-Control	Ireland, UK	Histologically proven AAV	Geographically matched healthy subjects	41/700	GPA (all patients)	NR	2.38 (0.70 – 8.12)	
	Nakamaru et al. 1996 ²⁰	Case-Control	Japan	Clinical, histological, serological criteria	Geographically matched healthy subjects	16/472	GPA (all patients)	c-ANCA (all patients)	1.71 (0.58 – 4.99)	

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Article	Study details		Definitions		Participants (n)		OR (95% CI)	
		Design	Setting	Cases	Controls	Cases / Controls	Diagnoses		ANCA serotype
<i>HLA-Cw1</i>	Nakamaru et al. 1996 ²⁰	Case-Control	Japan	Clinical, histological, serological criteria	Geographically matched healthy subjects	16/472	GPA (all patients)	c-ANCA (all patients)	1.45 (0.58 – 3.58)
	Von Vietinghoff et al. 2006 ¹⁸	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	52/51	GPA (all patients)	NR	0.98 (0.24 – 4.03)
	Nakamaru et al. 1996 ²⁰	Case-Control	Japan	Clinical, histological, serological criteria	Geographically matched healthy subjects	16/472	GPA (all patients)	c-ANCA (all patients)	1.40 (0.65 – 3.01)
<i>HLA-Cw7</i>	Von Vietinghoff et al. 2006 ¹⁸	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	52/51	GPA (all patients)	NR	1.32 (0.62 – 2.81)
	Nakamaru et al. 1996 ²⁰	Case-Control	Japan	Clinical, histological, serological criteria	Geographically matched healthy subjects	16/472	GPA (all patients)	c-ANCA (all patients)	0.50 (0.12 – 2.10)
	Von Vietinghoff et al. 2006 ¹⁸	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	52/51	GPA (all patients)	NR	1.10 (0.62 – 1.96)
<i>HLA-DPA1 rs9277341</i> (C)	Heckmann et al. 2008 ²⁴	Case-Control	Germany	ACR criteria	Geographically matched healthy blood donors	282/380	GPA (all patients)	ANCA+ (250), ANCA- (27)	0.38 (0.29 – 0.51)
	Xie et al. 2013 ^{3,5*}	Case-control (GWAS)	Canada, USA	ACR criteria	Geographically matched healthy subjects	459/1503	GPA (all patients)	c-ANCA (351)	0.30 (0.24 – 0.37)
<i>HLA-DPB1*0101</i>	Zhang et al. 1995 ²⁶	Case-Control	UK	CHCC criteria	Geographically matched healthy subjects	94/50	GPA (56), MPA (31), RVL (7)	PR3-ANCA (36), MPO-ANCA (19), PR3- and MPO-AN-CA positive (3)	1.20 (0.53 – 2.75)

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Article	Study details			Definitions		Participants (n)			OR (95% CI)
		Design	Setting	Cases	Controls	Cases / Controls	Diagnoses	ANCA serotype	Allele level	
HLA-DPB1*0201	Jagiello et al. 2004 ²⁷	Case-Control	Germany	ACR and CHCC criteria, biopsy	Geographically matched subjects	148/89	GPA (all patients)	PR3-ANCA (all patients)	0.17 (0.05 – 0.63)	
	Jagiello et al. 2004 ²⁷	Case-Control	Germany	ACR and CHCC criteria, biopsy	Geographically matched subjects	148/89	GPA (all patients)	PR3-ANCA (all patients)	0.78 (0.49 – 1.23)	
	Tsuchiya et al. 2006 ²⁸	Case-Control	Japan	Japanese 1998 criteria	Geographically matched healthy subjects	50/77	MPA (all patients)	MPO-ANCA (all patients)	0.44 (0.21 – 0.91)	
	Heckmann et al. 2008 ²⁴	Case-Control	Germany	ACR criteria	Geographically matched healthy blood donors	135/369	GPA (all patients)	ANCA+ (108), ANCA- (27)	1.38 (0.75 – 2.52)	
	Wieczorek et al. 2008 ²⁹	Case-Control	Germany	ACR criteria	Geographically matched healthy subjects	102/369	EGPA (all patients)	NR	0.93 (0.57 – 1.51)	
HLA-DPB1*0301	Zhang et al. 1995 ²⁶	Case-Control	UK	CHCC criteria	Geographically matched healthy subjects	94/50	GPA (56), MPA (31), RVL (7)	PR3-ANCA (36), MPO-ANCA (19), PR3- and MPO-AN-CA positive (3)	1.27 (0.56 – 2.89)	
	Jagiello et al. 2004 ²⁷	Case-Control	Germany	ACR and CHCC criteria, biopsy	Geographically matched subjects	148/89	GPA (all patients)	PR3-ANCA (all patients)	0.02 (0.00 – 0.15)	
	Heckmann et al. 2008 ²⁴	Case-Control	Germany	ACR criteria	Geographically matched healthy blood donors	135/369	GPA (all patients)	ANCA+ (108), ANCA- (27)	0.28 (0.14 – 0.55)	
	Wieczorek et al. 2008 ²⁹	Case-Control	Germany	ACR criteria	Geographically matched healthy subjects	102/369	EGPA (all patients)	NR	0.65 (0.38 – 1.11)	
	Arning et al. 2011 ^{10**}	Case-Control	Germany	CHCC criteria	Geographically and ethnically matched healthy blood donors	482/356	GPA (389), EGPA (56), MPA (37)	NR	0.30 (0.21 – 0.44)	
		Case-Control	UK	CHCC criteria	Geographically and ethnically matched healthy blood donors	193/104	GPA (102), MPA (82), EGPA (9)	NR	0.35 (0.16 – 0.77)	

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Article	Study details		Definitions		Cases / Controls		Participants (n)		OR (95% CI)
		Design	Setting	Cases	Controls	Cases / Controls	Diagnoses	ANCA serotype	Allele level	
HLA-DPBJ*0401	Zhang et al. 1995 ²⁶	Case-Control	UK	CHCC criteria	Geographically matched healthy subjects	94/50	GPA (56), MPA (31), RVL (7)	PR3-ANCA (36), MPO-ANCA (19), PR3- and MPO-AN-CA positive (3)	1.42 (0.84 – 2.38)	
	Jagiello et al. 2004 ²⁷	Case-Control	Germany	ACR and CHCC criteria, biopsy	Geographically matched subjects	148/89	GPA (all patients)	PR3-ANCA (all patients)	3.91 (2.62 – 5.84)	
	Heckmann et al. 2008 ²⁴	Case-Control	Germany	ACR criteria	Geographically matched healthy blood donors	135/369	GPA (all patients)	ANCA+ (108), ANCA- (27)	2.15 (1.61 – 2.86)	
	Wieczorek et al. 2008 ²⁹	Case-Control	Germany	ACR criteria	Geographically matched healthy subjects	102/369	EGPA (all patients)	NR	1.06 (0.78 – 1.45)	
	Arning et al. 2011 ^{30**}	Case-Control	Germany	CHCC criteria	Geographically and ethnically matched healthy blood donors	482/356	GPA (389), EGPA (56), MPA (37)	NR	2.34 (1.92 – 2.85)	
		Case-Control	UK	CHCC criteria	Geographically and ethnically matched healthy blood donors	193/104	GPA (102), MPA (82), EGPA (9)	NR	2.02 (1.44 – 2.85)	
HLA-DPBJ*0402	Zhang et al. 1995 ²⁶	Case-Control	UK	CHCC criteria	Geographically matched healthy subjects	94/50	GPA (56), MPA (31), RVL (7)	PR3-ANCA (36), MPO-ANCA (19), PR3- and MPO-AN-CA positive (3)	0.82 (0.38 – 1.78)	
	Jagiello et al. 2004 ²⁷	Case-Control	Germany	ACR and CHCC criteria, biopsy	Geographically matched subjects	148/89	GPA (all patients)	PR3-ANCA (all patients)	1.11 (0.65 – 1.92)	
	Heckmann et al. 2008 ²⁴	Case-Control	Germany	ACR criteria	Geographically matched healthy blood donors	135/369	GPA (all patients)	ANCA+ (108), ANCA- (27)	1.14 (0.78 – 1.68)	
	Wieczorek et al. 2008 ²⁹	Case-Control	Germany	ACR criteria	Geographically matched healthy subjects	102/369	EGPA (all patients)	NR	1.15 (0.75 – 1.77)	

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Article	Study details			Definitions		Participants (n)		OR (95% CI)
		Design	Setting	Cases	Controls	Cases / Controls	Diagnoses	ANCA serotype	
HLA-DPB1*0501	Zhang et al. 1995 ²⁶	Case-Control	UK	CHCC criteria	Geographically matched healthy subjects	94/50	GPA (56), MPA (31), RVL (7)	PR3-ANCA (36), MPO-ANCA (19), PR3- and MPO-AN-CA positive (3)	1.34 (0.26 – 7.02)
HLA-DPB1*0601	Jagiello et al. 2004 ²⁷	Case-Control	Germany	ACR and CHCC criteria, biopsy	Geographically matched subjects	148/89	GPA (all patients)	PR3-ANCA (all patients)	0.20 (0.01 – 4.93)
	Zhang et al. 1995 ²⁶	Case-Control	UK	CHCC criteria	Geographically matched healthy subjects	94/50	GPA (56), MPA (31), RVL (7)	PR3-ANCA (36), MPO-ANCA (19), PR3- and MPO-AN-CA positive (3)	3.79 (0.19 – 74.15)
HLA-DPB1*0901	Jagiello et al. 2004 ²⁷	Case-Control	Germany	ACR and CHCC criteria, biopsy	Geographically matched subjects	148/89	GPA (all patients)	PR3-ANCA (all patients)	0.60 (0.08 – 4.29)
	Zhang et al. 1995 ²⁶	Case-Control	UK	CHCC criteria	Geographically matched healthy subjects	94/50	GPA (56), MPA (31), RVL (7)	PR3-ANCA (36), MPO-ANCA (19), PR3- and MPO-AN-CA positive (3)	0.52 (0.15 – 1.84)
HLA-DPB2	Jagiello et al. 2004 ²⁷	Case-Control	Germany	ACR and CHCC criteria, biopsy	Geographically matched subjects	148/89	GPA (all patients)	PR3-ANCA (all patients)	1.81 (0.07 – 44.73)
rs3130215	Heckmann et al. 2008 ²⁴	Case-Control	Germany	ACR criteria	Geographically matched healthy blood donors	282/380	GPA (all patients)	ANCA+ (250), ANCA- (27)	2.35 (1.88 – 2.94)
(A)	Previously unpublished Lyons et al. 2012	Case-control (GWAS)	UK	EMA algorithm, supported by either serology or histology	Wellcome Trust Case Control Consortium (UK)	676/5365	GPA (394), MPA (156)	PR3-ANCA (326), MPO-ANCA (167)	0.88 (0.79 – 0.99)

GWAS data³

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Article	Study details		Definitions		Participants (n)		OR (95% CI)	
		Design	Setting	Cases	Controls	Cases / Controls	Diagnoses	ANCA serotype	Allele level
HLA-DQB1*02	Xie et al. 2013 ²⁵	Case-control (GWAS)	Canada, USA	ACR criteria	Geographically matched healthy subjects	459/1503	GPA (all patients)	c-ANCA (351)	2.44 (2.10 – 2.84)
	Spriewald et al. 2005 ³¹	Case-Control	Germany	ACR and CHCC criteria	Ethnically matched healthy subjects	32/91	GPA (all patients)	PR3-ANCA (all patients)	0.98 (0.45 – 2.13)
	Von Vietinghoff et al. 2006 ¹⁸	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	51/50	GPA (all patients)	NR	0.85 (0.41 – 1.77)
HLA-DQB1*0302	Zhang et al. 1995 ²⁶	Case-Control	UK	CHCC criteria	Geographically matched healthy subjects	94/50	GPA (56), MPA (31), RVL (7)	PR3-ANCA (36), MPO-ANCA (19), PR3- and MPO-AN-CA positive (3)	1.07 (0.53 – 2.20)
	Spriewald et al. 2005 ³¹	Case-Control	Germany	ACR and CHCC criteria	Ethnically matched healthy subjects	32/91	GPA (all patients)	PR3-ANCA (all patients)	0.94 (0.36 – 2.49)
HLA-DQB1*0303	Zhang et al. 1995 ²⁶	Case-Control	UK	CHCC criteria	Geographically matched healthy subjects	94/50	GPA (56), MPA (31), RVL (7)	PR3-ANCA (36), MPO-ANCA (19), PR3- and MPO-AN-CA positive (3)	0.84 (0.27 – 2.65)
	Spriewald et al. 2005 ³¹	Case-Control	Germany	ACR and CHCC criteria	Ethnically matched healthy subjects	32/91	GPA (all patients)	PR3-ANCA (all patients)	2.97 (0.72 – 12.23)
HLA-DQB1*04	Tsuchiya et al. 2006 ²⁸	Case-Control	Japan	Japanese 1998 criteria	Geographically matched healthy subjects	50/77	MPA (all patients)	MPO-ANCA (all patients)	2.11 (1.14 – 3.90)
	Spriewald et al. 2005 ³¹	Case-Control	Germany	ACR and CHCC criteria	Ethnically matched healthy subjects	32/91	GPA (all patients)	PR3-ANCA (all patients)	1.74 (0.40 – 7.50)
	Von Vietinghoff et al. 2006 ¹⁸	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	51/50	GPA (all patients)	NR	1.49 (0.24 – 9.08)

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Article	Study details			Definitions		Participants (n)			OR (95% CI)
		Design	Setting	Cases	Controls	Cases / Controls	Diagnoses	ANCA serotype	Allele level	
HLA-DQB1*0501	Zhang et al. 1995 ²⁶	Case-Control	UK	CHCC criteria	Geographically matched healthy subjects	94/50	GPA (56), MPA (31), RVL (7)	PR3-ANCA (36), MPO-ANCA (19), PR3- and MPO-AN-CA positive (3)	1.77 (0.91 – 3.42)	
	Spriewald et al. 2005 ³¹	Case-Control	Germany	ACR and CHCC criteria	Ethnically matched healthy subjects	32/91	GPA (all patients)	PR3-ANCA (all patients)	0.77 (0.32 – 1.88)	
HLA-DQB1*0602	Zhang et al. 1995 ²⁶	Case-Control	UK	CHCC criteria	Geographically matched healthy subjects	94/50	GPA (56), MPA (31), RVL (7)	PR3-ANCA (36), MPO-ANCA (19), PR3- and MPO-AN-CA positive (3)	0.64 (0.29 – 1.42)	
	Spriewald et al. 2005 ³¹	Case-Control	Germany	ACR and CHCC criteria	Ethnically matched healthy subjects	32/91	GPA (all patients)	PR3-ANCA (all patients)	12.86 (2.65 – 62.31)	
HLA-DQZ	Zhang et al. 1995 ²⁶	Case-Control	UK	CHCC criteria	Geographically matched healthy subjects	94/50	GPA (56), MPA (31), RVL (7)	PR3-ANCA (36), MPO-ANCA (19), PR3- and MPO-AN-CA positive (3)	1.21 (0.70 – 2.10)	
	Papasteriades et al. 1997 ²²	Case-Control	Greece	Histologically proven AAV	Geographically and ethnically matched healthy subjects	23/405	MPA (all patients)	NR	0.66 (0.34 – 1.30)	
HLA-DR1	Spriewald et al. 2005 ³¹	Case-Control	Germany	ACR and CHCC criteria	Ethnically matched healthy subjects	32/91	GPA (all patients)	PR3-ANCA (all patients)	0.43 (0.16 – 1.16)	
	Elkon et al. 1983 ²³	Case-Control	UK	Clinical and histological criteria	Geographically matched subjects	46/113	GPA (17), EGPA (14), PAN (15)	NR	2.17 (0.90 – 5.23)	
	Papiha et al. 1992 ³²	Case-Control	UK	Clinical and histological criteria	Geographically matched subjects	27/105	GPA (all patients)	NR	4.37 (2.01 – 9.48)	

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Study details		Definitions		Participants (n)		OR (95% CI)		
	Article	Design	Setting	Cases	Controls	Cases / Controls		Diagnoses	ANCA serotype
	Zhang et al. ¹⁹⁹⁵ ²⁶	Case-Control	UK	CHCC criteria	Geographically matched healthy subjects	94/90	GPA (56), MPA (31), RVL (7)	PR3-ANCA (36), MPO-ANCA (19), PR3- and MPO-AN-CA positive (3)	0.89 (0.44 – 1.82)
	Tsuchiya et al. ²⁰⁰³ ⁵	Case-Control	Japan	CHCC and Japanese 1998 criteria	Geographically matched healthy subjects	64/265	GPA (3), MPA (50), EGPA (8), PAN (3)	MPO-ANCA (all patients)	1.12 (0.48 – 2.64)
	Spriewald et al. ²⁰⁰⁵ ³¹	Case-Control	Germany	ACR and CHCC criteria	Ethnically matched healthy subjects	32/91	GPA (all patients)	PR3-ANCA (all patients)	0.68 (0.67 – 1.75)
	Von Vietinghoff et al. ²⁰⁰⁶ ¹⁸	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	51/51	GPA (all patients)	NR	0.56 (0.23 – 1.35)
	Wieczorek et al. ²⁰⁰⁸ ²⁹	Case-Control	Germany	ACR criteria	Geographically matched healthy subjects	102/369	EGPA (all patients)	NR	0.71 (0.43 – 1.15)
	Stassen et al. ²⁰⁰⁹ ¹⁹	Case-Control	The Netherlands	CHCC criteria	Geographically matched healthy blood donors	304/5442	GPA (241), MPA (30), EGPA (12), RVL (21)	PR3-ANCA (218), MPO-ANCA (64), other (4), ANCA negative (18)	0.67 (0.49 – 0.92)
	Cao et al. ²⁰¹¹ ³³	Case-Control	USA (96 Caucasians and 41 African Americans)	Histologically proven AAV	Ethnically matched community-based control frequencies (Healthy Organ Donor- North Carolina Services)	137/379	NR	PR3-ANCA (90), MPO-ANCA (47)	0.53 (0.34 – 0.81)
	Luo et al. ²⁰¹¹ ³⁴	Case-Control	China	EMA algorithm	Ethnically matched healthy blood donors	152/200	GPA (45), MPA (107)	PR3-ANCA (19), MPO-ANCA (130), PR3- and MPO-AN-CA positive (3)	0.79 (0.28 – 2.19)

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Article	Study details			Definitions		Participants (n)			OR (95% CI)
		Design	Setting	Cases	Controls	Cases / Controls	Diagnoses	ANCA serotype	Allele level	
HLA-DR2	Elkon et al. 1983 ²³	Case-Control	UK	Clinical and histological criteria	Geographically matched subjects	46/113	GPA (17), EGPA (14), PAN (15)	NR	1.91 (0.97 – 3.75)	
	Papiha et al. 1992 ³²	Case-Control	UK	Clinical and histological criteria	Geographically matched subjects	27/105	GPA (all patients)	NR	1.20 (0.56 – 2.54)	
	Zhang et al. 1995 ²⁶	Case-Control	UK	CHCC criteria	Geographically matched healthy subjects	94/90	GPA (56), MPA (31), RVL (7)	PR3-ANCA (36), MPO-ANCA (19), PR3- and MPO-AN-CA positive (3)	1.19 (0.66 – 2.13)	
	Nakamaru et al. 1996 ²⁰	Case-Control	Japan	Clinical, histological, serological criteria	Geographically matched healthy subjects	16/472	GPA (all patients)	c-ANCA (all patients)	0.33 (0.08 – 1.38)	
HLA-DR3	Stassen et al. 2009 ¹⁹	Case-Control	The Netherlands	CHCC criteria	Geographically matched healthy blood donors	304/5442	GPA (241), MPA (30), EGPA (12), RVL (21)	PR3-ANCA (218), MPO-ANCA (64), other (4), ANCA negative (18)	0.97 (0.77 – 1.22)	
	Elkon et al. 1983 ²³	Case-Control	UK	Clinical, and histological, criteria	Geographically matched subjects	46/113	GPA (17), EGPA (14), PAN (15)	NR	1.01 (0.52 – 1.99)	
	Papiha et al. 1992 ³²	Case-Control	UK	Clinical and histological criteria	Geographically matched subjects	27/105	GPA (all patients)	NR	1.41 (0.59 – 3.36)	
	Zhang et al. 1995 ²⁶	Case-Control	UK	CHCC criteria	Geographically matched healthy subjects	94/90	GPA (56), MPA (31), RVL (7)	PR3-ANCA (36), MPO-ANCA (19), PR3- and MPO-AN-CA positive (3)	0.75 (0.43 – 1.30)	
Papasteriades et al. 1997 ²²	Case-Control	Greece	Histologically proven AAV	Geographically and ethnically matched healthy subjects	23/405	MPA (all patients)	NR	0.16 (0.02 – 1.18)		

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Article		Study details		Definitions		Cases / Controls		Participants (n)		OR (95% CI)	
	Article	Design	Setting	Cases	Controls	Cases / Controls	Diagnoses	ANCA serotype	Allele level			
	Spriewald et al. 2005 ³¹	Case-Control	Germany	ACR and CHCC criteria	Ethnically matched healthy subjects	32/91	GPA (all patients)	PR3-ANCA (all patients)	2.54 (1.00 – 6.46)			
	Von Vietinghoff et al. 2006 ¹⁸	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	51/51	GPA (all patients)	NR	1.55 (0.53 – 4.52)			
	Wiczorek et al. 2008 ²⁹	Case-Control	Germany	ACR criteria	Geographically matched healthy subjects	102/369	EGPA (all patients)	NR	0.66 (0.39 – 1.11)			
	Stassen et al. 2009 ¹⁹	Case-Control	The Netherlands	CHCC criteria	Geographically matched healthy blood donors	304/5442	GPA (2+), MPA (30), EGPA (12), RVL (21)	PR3-ANCA (218), MPO-ANCA (6+), other (4), ANCA negative (18)	1.07 (0.84 – 1.35)			
	Cao et al. 2011 ³³	Case-Control	USA (96 Caucasians and 41 African Americans)	Histologically proven AAV	Ethnically matched community-based control frequencies (Healthy Organ Donor North Carolina Services)	137/379	NR	PR3-ANCA (90), MPO-ANCA (47)	1.30 (0.86 – 1.95)			
HLA-DR4	Elkon et al. 1983 ²³	Case-Control	UK	Clinical and histological criteria	Geographically matched subjects	46/113	GPA (17), EGPA (14), PAN (15)	NR	1.00 (0.52 – 1.92)			
	Murty et al. 1991 ¹⁷	Case-Control	Ireland, UK	Histologically proven AAV	Geographically matched healthy subjects	41/300	GPA (all patients)	NR	0.96 (0.50 – 1.83)			
	Papiha et al. 1992 ³²	Case-Control	UK	Clinical and histological criteria	Geographically matched subjects	27/105	GPA (all patients)	NR	0.81 (0.32 – 2.07)			

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Article	Study details		Definitions		Participants (n)			OR (95% CI)
		Design	Setting	Cases	Controls	Cases / Controls	Diagnoses	ANCA serotype	
	Zhang et al. ¹⁹⁹⁵ ²⁶	Case-Control	UK	CHCC criteria	Geographically matched healthy subjects	94/90	GPA (56), MPA (31), RVL (7)	PR3-ANCA (36), MPO-ANCA (19), PR3- and MPO-AN-CA positive (3)	1.24 (0.74 – 2.07)
	Nakamaru et al. ¹⁹⁹⁶ ²⁰	Case-Control	Japan	Clinical, histological, serological criteria	Geographically matched healthy subjects	16/472	GPA (all patients)	c-ANCA (all patients)	0.72 (0.27 – 1.88)
	Spriewald et al. ²⁰⁰⁵ ³¹	Case-Control	Germany	ACR and CHCC criteria	Ethnically matched healthy subjects	32/91	GPA (all patients)	PR3 (all patients)	1.25 (0.54 – 2.90)
	Von Vietinghoff et al. ²⁰⁰⁶ ¹⁸	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	51/51	GPA (all patients)	NR	1.57 (0.61 – 4.01)
	Vaglio et al. ²⁰⁰⁷ ³⁵	Case-Control	Italy	ACR and CHCC criteria	Geographically matched healthy subjects with no (family) history of autoimmune disease	48/350	EGPA (all patients)	MPO-ANCA (18), ANCA negative (26), undetermined (4)	1.73 (0.84 – 3.57)
	Wieczorek et al. ²⁰⁰⁸ ²⁹	Case-Control	Germany	ACR criteria	Geographically matched healthy subjects	102/341	EGPA (all patients)	NR	1.87 (1.23 – 2.82)
	Stassen et al. ²⁰⁰⁹ ¹⁹	Case-Control	The Netherlands	CHCC criteria	Geographically matched healthy blood donors	304/5442	GPA (241), MPA (30), EGPA (12), RVL (21)	PR3-ANCA (218), MPO-ANCA (64), other (4), ANCA negative (18)	1.53 (1.25 – 1.87)

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Article	Study details		Definitions		Cases / Controls	Participants (n)		OR (95% CI)
		Design	Setting	Cases	Controls		Diagnoses	ANCA serotype	
HLA-DR5	Cao et al. 2011 ^{33**}	Case-Control	USA (96 Caucasians and 41 African Americans)	Histologically proven AAV	Ethnically matched community-based control frequencies (Healthy Organ Donor- North Carolina Services)	137/379	NR	PR3-ANCA (90), MPO-ANCA (47)	0.73 (0.51 – 1.05)
		Case-Control	USA (all African Americans)	Histologically proven AAV	USA African Americans-Bethesda database	16/112	NR	PR3-ANCA (9), MPO-ANCA (4), p-ANCA (2), ANCA negative (1)	0.62 (0.08 – 5.01)
	Elkon et al. 1983 ²³	Case-Control	UK	Clinical and histological criteria	Geographically matched subjects	46/113	GPA (17), EGPA (14), PAN (15)	NR	0.72 (0.28 – 1.85)
	Papila et al. 1992 ³²	Case-Control	UK	Clinical and histological criteria	Geographically matched subjects	27/105	GPA (all patients)	NR	0.18 (0.02 – 1.37)
	Zhang et al. 1995 ²⁶	Case-Control	UK	CHCC criteria	Geographically matched healthy subjects	94/90	GPA (56), MPA (31), RVL (7)	PR3-ANCA (36), MPO-ANCA (19), PR3- and MPO-ANCA positive (3)	0.95 (0.45 – 2.01)
	Papasteriades et al. 1997 ²²	Case-Control	Greece	Histologically proven AAV	Geographically and ethnically matched healthy subjects	23/405	MPA (all patients)	NR	1.59 (0.80 – 3.15)
HLA-DR6	Stassen et al. 2009 ¹⁹	Case-Control	The Netherlands	CHCC criteria	Geographically matched healthy blood donors	304/5442	GPA (241), MPA (30), EGPA (12), RVL (21)	PR3-ANCA (218), MPO-ANCA (64), other (4), ANCA negative (18)	0.80 (0.58 – 1.09)
	Elkon et al. 1983 ²³	Case-Control	UK	Clinical and histological criteria	Geographically matched subjects	46/113	GPA (17), EGPA (14), PAN (15)	NR	0.16 (0.04 – 0.70)

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Article	Study details		Definitions		Controls		Participants (n)		OR (95% CI)
		Design	Setting	Cases	Controls	Cases / Controls	Diagnoses	ANCA serotype	Allele level	
	Papiha et al. 1992 ³²	Case-Control	UK	Clinical and histological criteria	Geographically matched subjects	27/105	GPA (all patients)	NR	0.11 (0.01 – 1.83)	
	Zhang et al. 1995 ³⁶	Case-Control	UK	CHCC criteria	Geographically matched healthy subjects	94/90	GPA (56), MPA (31), RVL (7)	PR3-ANCA (36), MPO-ANCA (19), PR3- and MPO-AN-CA positive (3)	0.60 (0.27 – 1.31)	
	Nakamaru et al. 1996 ²⁰	Case-Control	Japan	Clinical, histological, serological criteria	Geographically matched healthy subjects	16/472	GPA (all patients)	c-ANCA (all patients)	1.63 (0.56 – 4.77)	
	Stassen et al. 2009 ¹⁹	Case-Control	The Netherlands	CHCC criteria	Geographically matched healthy blood donors	304/5442	GPA (241), MPA (30), EGPA (12), RVL (21)	PR3-ANCA (218), MPO-ANCA (64), other (4), ANCA negative (18)	0.44 (0.32 – 0.59)	
HLA-DR7	Elkon et al. 1983 ²³	Case-Control	UK	Clinical and histological criteria	Geographically matched subjects	46/113	GPA (17), EGPA (14), PAN (15)	NR	0.512 (0.22 – 1.22)	
	Papiha et al. 1992 ³²	Case-Control	UK	Clinical and histological criteria	Geographically matched subjects	27/105	GPA (all patients)	NR	0.64 (0.23 – 1.73)	
	Spriewald et al. 2005 ³¹	Case-Control	Germany	ACR and CHCC criteria	Ethnically matched healthy subjects	32/91	GPA (all patients)	PR3 (all patients)	0.99 (0.40 – 2.48)	
	Von Vietinghoff et al. 2006 ¹⁸	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	51/51	GPA (all patients)	NR	0.77 (0.34 – 1.75)	
	Vaglio et al. 2007 ³⁵	Case-Control	Italy	ACR and CHCC criteria	Geographically matched healthy subjects with no (family) history of autoimmune disease	48/350	EGPA (all patients)	MPO-ANCA (18), ANCA negative (26), undetermined (4)	2.42 (1.47 – 4.00)	

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Article		Study details		Definitions		Cases / Controls	Participants (n)		OR (95% CI)
	Article	Design	Setting	Cases	Controls	Diagnoses		ANCA serotype	Allele level	
	Wieczorek et al. 2008 ²⁹	Case-Control	Germany	ACR criteria	Geographically matched healthy subjects	102/341	EGPA (all patients)	NR	1.58 (1.01 – 2.47)	
	Stassen et al. 2009 ¹⁹	Case-Control	The Netherlands	CHCC criteria	Geographically matched healthy blood donors	304/5442	GPA (241), MPA (30), EGPA (12), RVL (21)	PR3-ANCA (218), MPO-ANCA (64), other (4), ANCA negative (18)	0.96 (0.72 – 1.27)	
HLA-DR8	Elkon et al. 1983 ²³	Case-Control	UK	Clinical and histological criteria	Geographically matched subjects	46/113	GPA (17), EGPA (14), PAN (15)	NR	0.52 (0.22 – 1.22)	
	Murty et al. 1991 ¹⁷	Case-Control	Ireland, UK	Histologically proven AAV	Geographically matched healthy subjects	41/300	GPA (all patients)	NR	1.05 (0.21 – 5.14)	
	Zhang et al. 1995 ²⁶	Case-Control	UK	CHCC criteria	Geographically matched healthy subjects	94/90	GPA (56), MPA (31), RVL (7)	PR3-ANCA (36), MPO-ANCA (19), PR3- and MPO-AN-CA positive (3)	2.21 (0.67 – 7.32)	
	Nakamaru et al. 1996 ²⁰	Case-Control	Japan	Clinical, histological, serological criteria	Geographically matched healthy subjects	16/472	GPA (all patients)	c-ANCA (all patients)	1.63 (0.66 – 4.05)	
	Spriewald et al. 2005 ³¹	Case-Control	Germany	ACR and CHCC criteria	Ethnically matched healthy subjects	32/91	GPA (all patients)	PR3 (all patients)	1.23 (0.31 – 4.91)	
	Von Vietinghoff et al. 2006 ¹⁸	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	51/51	GPA (all patients)	NR	1.00 (0.12 – 5.08)	
	Wieczorek et al. 2008 ²⁹	Case-Control	Germany	ACR criteria	Geographically matched healthy subjects	102/369	EGPA (all patients)	NR	2.03 (1.01 – 4.05)	

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Article	Study details			Definitions		Participants (n)			OR (95% CI)
		Design	Setting	Cases	Controls	Cases / Controls	Diagnoses	ANCA serotype	Allele level	
	Stassen et al. 2009 ¹⁹	Case-Control	UK	CHCC criteria	Geographically matched healthy subjects	94/90	GPA (56), MPA (31), RVL (7)	PR3-ANCA (36), MPO-ANCA (19), PR3- and MPO-ANCA positive (3)	0.95 (0.59 – 1.54)	
	Cao et al. 2011 ³³	Case-Control	USA (96 Caucasians and 41 African Americans)	Histologically proven AAV	Ethnically matched community-based control frequencies (Healthy Organ Donors North Carolina Services)	137/379	NR	PR3-ANCA (90), MPO-ANCA (47)	0.76 (0.37 – 1.55)	
HLA-DR9	Elkon et al. 1983 ²³	Case-Control	UK	Clinical and histological criteria	Geographically matched subjects	46/113	GPA (17), EGPA (14), PAN (15)	NR	0.49 (0.02 – 10.21)	
	Nakamaru et al. 1996 ²⁰	Case-Control	Japan	Clinical, histological, serological criteria	Geographically matched healthy subjects	16/472	GPA (all patients)	c-ANCA (all patients)	3.03 (1.40 – 6.56)	
	Fujii et al. 2000 ³⁶	Case-Control	Japan	CHCC criteria	Geographically matched healthy subjects	12/472	NR	MPO-ANCA (all patients)	2.75 (1.12 – 6.76)	
	Spriewald et al. 2005 ³¹	Case-Control	Germany	ACR and CHCC criteria	Ethnically matched healthy subjects	32/91	GPA (all patients)	PR3 (all patients)	0.94 (0.04 – 23.32)	
	Von Vietinghoff et al. 2006 ¹⁸	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	51/51	GPA (all patients)	NR	0.66 (0.11 – 4.04)	
	Vaglio et al. 2007 ³⁵	Case-Control	Italy	ACR and CHCC criteria	Geographically matched healthy subjects with no (family) history of autoimmune disease	48/350	EGPA (all patients)	MPO-ANCA (18), ANCA negative (26), undetermined (4)	1.83 (0.20 – 16.56)	
	Wieczorek et al. 2008 ²⁹	Case-Control	Germany	ACR criteria	Geographically matched healthy subjects	102/369	EGPA (all patients)	NR	0.80 (0.17 – 3.74)	

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Article	Study details		Definitions		Participants (n)		OR (95% CI)	
		Design	Setting	Cases	Controls	Cases / Controls	Diagnoses		ANCA serotype
	Stassen et al. 2009 ¹⁹	Case-Control	UK	CHCC criteria	Geographically matched healthy subjects	94/90	GPA (56), MPA (31), RVL (7)	PR3-ANCA (36), MPO-ANCA (19), PR3- and MPO-AN-CA positive (3)	0.35 (0.11 – 1.09)
	Cao et al. 2011 ³³	Case-Control	USA (96 Caucasians and 41 African Americans)	Histologically proven AAV	Ethnically matched community-based control frequencies (Healthy Organ Donor- North Carolina Services)	137/379	NR	PR3-ANCA (90), MPO-ANCA (47)	0.75 (0.208 – 2.72)
	Luo et al. 2011 ³⁴	Case-Control	China	EMA algorithm	Ethnically matched healthy blood donors	152/200	GPA (45), MPA (107)	PR3-ANCA (19), MPO-ANCA (130), PR3- and MPO-AN-CA positive (3)	0.68 (0.43 – 1.06)
	Tsuchiya et al. 2013 ³⁷	Case-Control	Japan	CHCC and Japanese 1998 criteria	Geographically matched healthy subjects	116/265	MPA (96)	MPO-ANCA (all patients)	1.74 (1.18 – 2.57)
HLA-DRB1*0401	Tsuchiya et al. 2003 ⁵	Case-Control	Japan	CHCC and Japanese 1998 criteria	Geographically matched healthy subjects	64/265	GPA (3), MPA (50), EGPA (8), PAN (3)	MPO-ANCA (all patients)	2.10 (0.52 – 8.50)
	Luo et al. 2011 ³⁴	Case-Control	China	EMA algorithm	Ethnically matched healthy blood donors	152/200	GPA (45), MPA (107)	PR3-ANCA (19), MPO-ANCA (130), PR3- and MPO-AN-CA positive (3)	0.58 (0.18 – 1.90)
HLA-DRB1*0403	Tsuchiya et al. 2003 ⁵	Case-Control	Japan	CHCC and Japanese 1998 criteria	Geographically matched healthy subjects	64/265	GPA (3), MPA (50), EGPA (8), PAN (3)	MPO-ANCA (all patients)	0.95 (0.27 – 3.40)

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Article	Study details			Definitions		Cases / Controls	Participants (n)		OR (95% CI)
		Design	Setting	Cases	Controls	Diagnoses		ANCA serotype	Allele level	
	Luo et al. 2011 ³⁴	Case-Control	China	EMA algorithm	Ethnically matched healthy blood donors	152/200	GPA (45), MPA (107)	PR3-ANCA (19), MPO-ANCA (130), PR3- and MPO-AN-CA positive (3)	0.16 (0.02 – 1.30)	
HLA-DRB1*0405	Tsuchiya et al. 2003 ⁵	Case-Control	Japan	CHCC and Japanese 1998 criteria	Geographically matched healthy subjects	64/265	GPA (3), MPA (50), EGPA (8), PAN (3)	MPO-ANCA (all patients)	0.61 (0.30 – 1.22)	
	Luo et al. 2011 ³⁴	Case-Control	China	EMA algorithm	Ethnically matched healthy blood donors	152/200	GPA (45), MPA (107)	PR3-ANCA (19), MPO-ANCA (130), PR3- and MPO-AN-CA positive (3)	1.13 (0.52 – 2.49)	
HLA-DRB1*0406	Tsuchiya et al. 2003 ⁵	Case-Control	Japan	CHCC and Japanese 1998 criteria	Geographically matched healthy subjects	64/265	GPA (3), MPA (50), EGPA (8), PAN (3)	MPO-ANCA (all patients)	0.65 (0.19 – 2.22)	
	Luo et al. 2011 ³⁴	Case-Control	China	EMA algorithm	Ethnically matched healthy blood donors	152/200	GPA (45), MPA (107)	PR3-ANCA (19), MPO-ANCA (130), PR3- and MPO-AN-CA positive (3)	1.84 (0.73 – 4.63)	
HLA-DRB1*0407	Tsuchiya et al. 2003 ⁵	Case-Control	Japan	CHCC and Japanese 1998 criteria	Geographically matched healthy subjects	64/265	GPA (3), MPA (50), EGPA (8), PAN (3)	MPO-ANCA (all patients)	2.82 (0.78 – 10.14)	
	Luo et al. 2011 ³⁴	Case-Control	China	EMA algorithm	Ethnically matched healthy blood donors	152/200	GPA (45), MPA (107)	PR3-ANCA (19), MPO-ANCA (130), PR3- and MPO-AN-CA positive (3)	0.44 (0.02 – 10.77)	

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Article	Study details		Definitions		Participants (n)		OR (95% CI)	
		Design	Setting	Cases	Controls	Cases / Controls	Diagnoses		ANCA serotype
HLA-DRB1*0410	Tsuchiya et al. 2003 ⁵	Case-Control	Japan	CHCC and Japanese 1998 criteria	Geographically matched healthy subjects	64/265	GPA (3), MPA (50), EGPA (8), PAN (3)	MPO-ANCA (all patients)	1.67 (0.32 – 8.69)
	Luo et al. 2011 ^{3*}	Case-Control	China	EMA algorithm	Ethnically matched healthy blood donors	152/200	GPA (45), MPA (107)	PR3-ANCA (19), MPO-ANCA (130), PR3- and MPO-AN-CA positive (3)	0.44 (0.02 – 10.77)
HLA-DRB1*0802	Tsuchiya et al. 2003 ⁵	Case-Control	Japan	CHCC and Japanese 1998 criteria	Geographically matched healthy subjects	64/265	GPA (3), MPA (50), EGPA (8), PAN (3)	MPO-ANCA (all patients)	1.16 (0.42 – 3.18)
	Luo et al. 2011 ^{3*}	Case-Control	China	EMA algorithm	Ethnically matched healthy blood donors	152/200	GPA (45), MPA (107)	PR3-ANCA (19), MPO-ANCA (130), PR3- and MPO-AN-CA positive (3)	0.26 (0.01 – 5.47)
HLA-DRB1*0803	Tsuchiya et al. 2003 ⁵	Case-Control	Japan	CHCC and Japanese 1998 criteria	Geographically matched healthy subjects	64/265	GPA (3), MPA (50), EGPA (8), PAN (3)	MPO-ANCA (all patients)	1.51 (0.78 – 2.93)
	Luo et al. 2011 ^{3*}	Case-Control	China	EMA algorithm	Ethnically matched healthy blood donors	152/200	GPA (45), MPA (107)	PR3-ANCA (19), MPO-ANCA (130), PR3- and MPO-AN-CA positive (3)	1.19 (0.61 – 2.33)
HLA-DRB1*10	Spriewald et al. 2005 ³¹	Case-Control	Germany	ACR and CHCC criteria	Ethnically matched healthy subjects	32/91	GPA (all patients)	PR3 (all patients)	1.43 (0.13 – 16.03)
	Von Vietinghoff et al. 2006 ¹⁸	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	51/51	GPA (all patients)	NR	5.10 (0.24 – 107.55)

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Article	Study details			Definitions		Participants (n)			OR (95% CI) Allele level
		Design	Setting	Cases	Controls	Cases / Controls	Diagnoses	ANCA serotype		
HLA- DRB1*11	Wiczorek et al. 2008 ²⁹	Case-Control	Germany	ACR criteria	Geographically matched healthy subjects	102/369	EGPA (all patients)	NR	0.40 (0.02 – 7.44)	
	Cao et al. 2011 ³³	Case-Control	USA (96 Caucasians and 41 African Americans)	Histologically proven AAV	Ethnically matched community-based control frequencies (Healthy Organ Donor-North Carolina Services)	137/379	NR	PR3-ANCA (90), MPO-ANCA (47)	0.75 (0.21 – 2.72)	
	Spriewald et al. 2005 ³¹	Case-Control	Germany	ACR and CHCC criteria	Ethnically matched healthy subjects	32/91	GPA (all patients)	PR3 (all patients)	0.723 (0.26 – 2.04)	
	Von Vietinghoff et al. 2006 ¹⁸	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	51/51	GPA (all patients)	NR	1.28 (0.48 – 3.38)	
HLA- DRB1*1101	Wiczorek et al. 2008 ²⁹	Case-Control	Germany	ACR criteria	Geographically matched healthy subjects	102/369	EGPA (all patients)	NR	0.88 (0.53 – 1.48)	
	Cao et al. 2011 ³³	Case-Control	USA (96 Caucasians and 41 African Americans)	Histologically proven AAV	Ethnically matched community-based control frequencies (Healthy Organ Donor-North Carolina Services)	137/379	NR	PR3-ANCA (90), MPO-ANCA (47)	1.09 (0.66 – 1.80)	
HLA- DRB1*1101	Luo et al. 2011 ³⁴	Case-Control	China	EMA algorithm	Ethnically matched healthy blood donors	152/200	GPA (45), MPA (107)	PR3-ANCA (19), MPO-ANCA (130), PR3- and MPO-AN-CA positive (3)	1.75 (1.02 – 3.00)	
	Tsuchiya et al. 2013 ³⁷	Case-Control	Japan	CHCC and Japanese 1998 criteria	Geographically matched healthy subjects	116/265	MPA (96)	MPO-ANCA (all patients)	2.79 (0.84 – 9.23)	
HLA- DRB1*12	Spriewald et al. 2005 ³¹	Case-Control	Germany	ACR and CHCC criteria	Ethnically matched healthy subjects	32/91	GPA (all patients)	PR3 (all patients)	2.97 (0.721 – 2.23)	

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Article	Study details		Definitions		Cases / Controls		Participants (n)		OR (95% CI)	
		Design	Setting	Cases	Controls	Cases / Controls	Diagnoses	ANCA serotype	Allele level		
<i>HLA-DRB1*1201</i>	Von Vietinghoff et al. 2006 ¹⁸	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	51/51	GPA (all patients)	NR	6.31 (0.75 – 53.40)		
	Wieczorek et al. 2008 ²⁹	Case-Control	Germany	ACR criteria	Geographically matched healthy subjects	102/369	EGPA (all patients)	NR	0.57 (0.17 – 1.93)		
	Cao et al. 2011 ³³	Case-Control	USA (96 Caucasians and 41 African Americans)	Histologically proven AAV	Ethnically matched community-based control frequencies (Healthy Organ Donor- North Carolina Services)	137/379	NR	PR3-ANCA (90), MPO-ANCA (47)	0.59 (0.17 – 2.06)		
<i>HLA-DRB1*1202</i>	Tsuchiya et al. 2003 ⁵	Case-Control	Japan	CHCC and Japanese 1998 criteria	Geographically matched healthy subjects	64/265	GPA (3), MPA (50), EGPA (8), PAN (3)	MPO-ANCA (all patients)	0.21 (0.03 – 1.60)		
	Luo et al. 2011 ³⁴	Case-Control	China	EMA algorithm	Ethnically matched healthy blood donors	152/200	GPA (45), MPA (107)	PR3-ANCA (19), MPO-ANCA (130), PR3- and MPO-AN-CA positive (3)	0.46 (0.16 – 1.29)		
	Tsuchiya et al. 2003 ⁵	Case-Control	Japan	CHCC and Japanese 1998 criteria	Geographically matched healthy subjects	64/265	GPA (3), MPA (50), EGPA (8), PAN (3)	MPO-ANCA (all patients)	0.37 (0.05 – 2.90)		
<i>HLA-DRB1*13</i>	Luo et al. 2011 ³⁴	Case-Control	China	EMA algorithm	Ethnically matched healthy blood donors	152/200	GPA (45), MPA (107)	PR3-ANCA (19), MPO-ANCA (130), PR3- and MPO-AN-CA positive (3)	1.70 (0.93 – 3.13)		
	Spriewald et al. 2005 ³¹	Case-Control	Germany	ACR and CHCC criteria	Ethnically matched healthy subjects	32/91	GPA (all patients)	PR3 (all patients)	0.56 (0.20 – 1.53)		

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Article	Study details			Definitions		Participants (n)			OR (95% CI)
		Design	Setting	Cases	Controls	Cases / Controls	Diagnoses	ANCA serotype	Allele level	
	Von Vietinghoff et al. 2006 ¹⁸	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	51/51	GPA (all patients)	NR	0.65 (0.31 – 1.38)	
	Vaglio et al. 2007 ³⁵	Case-Control	Italy	ACR and CHCC criteria	Geographically matched healthy subjects with no (family) history of autoimmune disease	48/350	EGPA (all patients)	MPO-ANCA (18), ANCA negative (26), undetermined (4)	0.23 (0.07 – 0.73)	
	Wieczorek et al. 2008 ²⁹	Case-Control	Germany	ACR criteria	Geographically matched healthy subjects	102/341	EGPA (all patients)	NR	0.50 (0.28 – 0.89)	
HLA-DRB1*1302	Tsuchiya et al. 2003 ⁵	Case-Control	Japan	CHCC and Japanese 1998 criteria	Geographically matched healthy subjects	64/265	GPA (3), MPA (50), EGPA (8), PAN (3)	MPO-ANCA (all patients)	0.54 (0.24 – 1.23)	
	Luo et al. 2011 ³⁴	Case-Control	China	EMA algorithm	Ethnically matched healthy blood donors	152/200	GPA (45), MPA (107)	PR3-ANCA (19), MPO-ANCA (130), PR3- and MPO-AN-CA positive (3)	0.52 (0.16 – 1.67)	
HLA-DRB1*14	Spriewald et al. 2005 ³¹	Case-Control	Germany	ACR and CHCC criteria	Ethnically matched healthy subjects	32/91	GPA (all patients)	PR3 (all patients)	1.07 (0.27 – 4.16)	
	Von Vietinghoff et al. 2006 ¹⁸	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	51/51	GPA (all patients)	NR	2.02 (0.18 – 22.63)	
	Wieczorek et al. 2008 ²⁹	Case-Control	Germany	ACR criteria	Geographically matched healthy subjects	102/341	EGPA (all patients)	NR	1.83 (0.72 – 4.65)	

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Article	Study details		Definitions		Cases / Controls	Participants (n)		OR (95% CI)
		Design	Setting	Cases	Controls		Diagnoses	ANCA serotype	
	Cao et al. 2011 ³³	Case-Control	USA (96 Caucasians and 41 African Americans)	Histologically proven AAV	Ethnically matched community-based control frequencies (Healthy Organ Donor-North Carolina Services)	137/379	NR	PR3-ANCA (90), MPO-ANCA (47)	2.82 (1.05 – 7.59)
HLA-DRB1*1403	Tsuchiya et al. 2003 ⁵	Case-Control	Japan	CHCC and Japanese 1998 criteria	Geographically matched healthy subjects	64/265	GPA (3), MPA (50), EGPA (8), PAN (3)	MPO-ANCA (all patients)	1.25 (0.34 – 4.60)
	Luo et al. 2011 ³⁴	Case-Control	China	EMA algorithm	Ethnically matched healthy blood donors	152/200	GPA (45), MPA (107)	PR3-ANCA (19), MPO-ANCA (130), PR3- and MPO-AN-CA positive (3)	1.32 (0.19 – 9.41)
HLA-DRB1*1405	Tsuchiya et al. 2003 ⁵	Case-Control	Japan	CHCC and Japanese 1998 criteria	Geographically matched healthy subjects	64/265	GPA (3), MPA (50), EGPA (8), PAN (3)	MPO-ANCA (all patients)	0.11 (0.01 – 1.91)
	Luo et al. 2011 ³⁴	Case-Control	China	EMA algorithm	Ethnically matched healthy blood donors	152/200	GPA (45), MPA (107)	PR3-ANCA (19), MPO-ANCA (130), PR3- and MPO-AN-CA positive (3)	0.96 (0.44 – 2.13)
HLA-DRB1*15	Spriewald et al. 2005 ³¹	Case-Control	Germany	ACR and CHCC criteria	Ethnically matched healthy subjects	32/91	GPA (all patients)	PR3 (all patients)	0.79 (0.34 – 1.83)
	Von Vietinghoff et al. 2006 ¹⁸	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	51/51	GPA (all patients)	NR	1.13 (0.57 – 2.26)

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Article	Study details			Definitions		Participants (n)			OR (95% CI)
		Design	Setting	Cases	Controls	Cases / Controls	Diagnoses	ANCA serotype	Allele level	
	Cao et al. 2011 ^{33**}	Case-Control	USA (96 Caucasians and 41 African Americans)	Histologically proven AAV	Ethnically matched community-based control frequencies (Healthy Organ Donors- North Carolina Services)	137/379	NR	PR3-ANCA (90), MPO-ANCA (47)	2.72 (1.84 – 4.03)	
	Case-Control	USA (all African Americans)	Histologically proven AAV	USA African Americans-Bethesda database	16/112	NR	PR3-ANCA (9), MPO-ANCA (4), p-ANCA (2), ANCA negative (1)	2.08 (0.86 – 5.03)		
HLA-DRB1*1501	Tsuchiya et al. 2003 ⁵	Case-Control	Japan	CHCC and Japanese 1998 criteria	Geographically matched healthy subjects	64/265	GPA (3), MPA (50), EGPA (8), PAN (3)	MPO-ANCA (all patients)	1.62 (0.79 – 3.35)	
	Case-Control	China	EMA algorithm	Ethnically matched healthy blood donors	152/200	GPA (45), MPA (107)	PR3-ANCA (19), MPO-ANCA (130), PR3- and MPO-ANCA positive (3)	1.69 (1.16 – 2.46)		
HLA-DRB1*1502	Tsuchiya et al. 2003 ⁵	Case-Control	Japan	CHCC and Japanese 1998 criteria	Geographically matched healthy subjects	64/265	GPA (3), MPA (50), EGPA (8), PAN (3)	MPO-ANCA (all patients)	0.67 (0.32 – 1.39)	
	Case-Control	China	EMA algorithm	Ethnically matched healthy blood donors	152/200	GPA (45), MPA (107)	PR3-ANCA (19), MPO-ANCA (130), PR3- and MPO-ANCA positive (3)	0.35 (0.10 – 1.27)		
HLA-DRB1*16	Spriewald et al. 2005 ³¹	Case-Control	Germany	ACR and CHCC criteria	Ethnically matched healthy subjects	32/91	GPA (all patients)	PR3 (all patients)	0.40 (0.02 – 7.80)	

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Article	Study details		Definitions		Cases / Controls		Participants (n)		OR (95% CI)
		Design	Setting	Cases	Controls	Cases / Controls	Diagnoses	ANCA serotype	Allele level	
<i>HLA-DRB1*1602</i>	Von Vietinghoff et al. 2006 ¹⁸	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	51/51	GPA (all patients)	NR	0.14 (0.01 – 2.72)	
	Cao et al. 2011 ^{33**}	Case-Control	USA (96 Caucasians and 41 African Americans)	Histologically proven AAV	Ethnically matched community-based control frequencies (Healthy Organ Donors North Carolina Services)	137/379	NR	PR3-ANCA (90), MPO-ANCA (47)	1.39 (0.34 – 5.59)	
		Case-Control	USA (all African Americans)	Histologically proven AAV	USA African Americans-Bethesda database	16/112	NR	PR3-ANCA (9), MPO-ANCA (4), p-ANCA (2), ANCA negative (1)	2.42 (0.47 – 12.55)	
<i>HLA-DRB1*1602</i>	Tsuchiya et al. 2003 ⁵	Case-Control	Japan	CHCC and Japanese 1998 criteria	Geographically matched healthy subjects	64/265	GPA (3), MPA (50), EGPA (8), PAN (3)	MPO-ANCA (all patients)	0.24 (0.01 – 4.171)	
	Luo et al. 2011 ³⁴	Case-Control	China	EMA algorithm	Ethnically matched healthy blood donors	152/200	GPA (45), MPA (107)	PR3-ANCA (19), MPO-ANCA (130), PR3- and MPO-ANCA positive (3)	0.12 (0.01 – 2.14)	
<i>HLA-DRB3</i>	Spencer et al. 1992 ³⁸	Case-Control	UK	Clinical criteria	Geographically and ethnically matched subjects	59/1103	GPA (34), MPA (25)	c-ANCA (47), p-ANCA (10), ANCA negative (2)	0.29 (0.11 – 0.79)	
<i>HLA-DRB3</i>	Von Vietinghoff et al. 2006 ¹⁸	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	51/51	GPA (all patients)	NR	1.41 (0.76 – 2.60)	
	Vaglio et al. 2007 ³⁵	Case-Control	Italy	ACR and CHCC criteria	Geographically matched healthy subjects with no (family) history of autoimmune disease	48/350	EGPA (all patients)	MPO-ANCA (18), ANCA negative (26), undetermined (4)	0.54 (0.35 – 0.84)	

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Article	Study details			Definitions		Participants (n)			OR (95% CI) Allele level
		Design	Setting	Cases	Controls	Cases / Controls	Diagnoses	ANCA serotype		
HLA-DRB4	Wieczorek et al. 2008 ²⁹	Case-Control	Germany	ACR criteria	Geographically matched healthy subjects	102/341	EGPA (all patients)	NR	0.61 (0.44 – 0.86)	
	Spencer et al. 1992 ³⁸	Case-Control	UK	Clinical criteria	Geographically and ethnically matched subjects	59/1103	GPA (34), MPA (25)	c-ANCA (47), p-ANCA (10), ANCA negative (2)	1.38 (0.89 – 2.13)	
	Von Vietinghoff et al. 2006 ¹⁸	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	51/51	GPA (all patients)	NR	0.89 (0.46 – 1.72)	
	Vaglio et al. 2007 ³⁵	Case-Control	Italy	ACR and CHCC criteria	Geographically matched healthy subjects with no (family) history of autoimmune disease	48/350	EGPA (all patients)	MPO-ANCA (18), ANCA negative (26), undetermined (4)	2.49 (1.58 – 3.90)	
HLA-DRB5	Wieczorek et al. 2008 ²⁹	Case-Control	Germany	ACR criteria	Geographically matched healthy subjects	102/341	EGPA (all patients)	NR	1.87 (1.34 – 2.61)	
	Spencer et al. 1992 ³⁸	Case-Control	UK	Clinical criteria	Geographically and ethnically matched subjects	59/1103	GPA (34), MPA (25)	c-ANCA (47), p-ANCA (10), ANCA negative (2)	1.35 (0.69 – 2.63)	
HSD17B8 rs421446 (C)	Von Vietinghoff et al. 2006 ¹⁸	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	51/51	GPA (all patients)	NR	1.07 (0.82 – 2.18)	
	Heckmann et al. 2008 ²⁴	Case-Control	Germany	ACR criteria	Geographically matched healthy blood donors	282/369	GPA (all patients)	ANCA+ (250), ANCA- (27)	0.43 (0.32 – 0.57)	
	Xie et al. 2013 ³⁵	Case-control (GWAS)	Canada, USA	ACR criteria	Geographically matched healthy subjects	459/1503	GPA (all patients)	c-ANCA (351)	0.39 (0.32 – 0.48)	
IL-1β (A2)	Huang et al. 2000 ⁴	Case-Control	Sweden	ACR criteria	Geographically and ethnically matched healthy subjects	32/109	GPA (all patients)	PR3-ANCA (29)	1.21 (0.55 – 2.63)	
	Borgmann et al. 2003 ³⁹	Case-Control	Germany	CHCC criteria	Geographically matched healthy, unrelated subjects	109/196	NR	PR3-ANCA (79), MPO-ANCA (30)	0.89 (0.61 – 1.29)	

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Article	Study details		Definitions		Cases / Controls		Participants (n)		OR (95% CI)
		Design	Setting	Cases	Controls	Cases / Controls	Diagnoses	ANCA serotype	Allele level	
	Zhou et al. 2004 ⁶	Case-Control	USA	ACR criteria	Geographically and ethnically matched healthy, unrelated subjects	117/123	GPA (all patients)	PR3-ANCA (82), MPO-ANCA (5), PR3- and MPO-ANCA positive (2), ANCA negative (25)	1.16 (0.72 – 1.85)	
	Previously unpublished Lyons et al. 2012 GWAS data ³	Case-control (GWAS)	UK	EMA algorithm, supported by either serology or histology	Wellcome Trust Case Control Consortium (UK)	914/5259	GPA (565), MPA (262)	PR3-ANCA (478), MPO-ANCA (264)	1.00 (0.89 – 1.12)	
ILIRN*1	Borgmann et al. 2003 ³⁹	Case-Control	Germany	CHCC criteria	Geographically matched healthy, unrelated subjects	109/234	NR	PR3-ANCA (79), MPO-ANCA (30)	1.12 (0.79 – 1.59)	
	Persson et al. 2013 ⁷	Case-Control	Sweden	ACR and CHCC criteria, EMA algorithm	Blood donors	105/200	GPA (61), MPA (44)	PR3-ANCA (62), MPO-ANCA (43)	1.00 (0.69 – 1.44)	
ILIRN*2	Borgmann et al. 2003 ³⁹	Case-Control	Germany	CHCC criteria	Geographically matched healthy, unrelated subjects	109/234	NR	PR3-ANCA (79), MPO-ANCA (30)	1.00 (0.69 – 1.43)	
	Persson et al. 2013 ⁷	Case-Control	Sweden	ACR and CHCC criteria, EMA algorithm	Blood donors	105/200	GPA (61), MPA (44)	PR3-ANCA (62), MPO-ANCA (43)	1.01 (0.69 – 1.48)	
ILIRN*3	Borgmann et al. 2003 ³⁹	Case-Control	Germany	CHCC criteria	Geographically matched healthy, unrelated subjects	109/234	NR	PR3-ANCA (79), MPO-ANCA (30)	0.30 (0.07 – 1.33)	
	Persson et al. 2013 ⁷	Case-Control	Sweden	ACR and CHCC criteria, EMA algorithm	Blood donors	105/200	GPA (61), MPA (44)	PR3-ANCA (62), MPO-ANCA (43)	1.06 (0.35 – 3.20)	
ILIRN*4	Borgmann et al. 2003 ³⁹	Case-Control	Germany	CHCC criteria	Geographically matched healthy, unrelated subjects	109/234	NR	PR3-ANCA (79), MPO-ANCA (30)	0.71 (0.03 – 17.58)	

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Article	Study details			Definitions		Participants (n)			OR (95% CI)
		Design	Setting	Cases	Controls	Cases / Controls	Diagnoses	ANCA serotype	Allele level	
	Persson et al. 2013 ⁷	Case-Control	Sweden	ACR and CHCC criteria, EMA algorithm	Blood donors	105/200	GPA (61), MPA (44)	PR3-ANCA (62), MPO-ANCA (43)	0.95 (0.09 – 10.56)	
IL1RN*5	Borgmann et al. 2003 ³⁹	Case-Control	Germany	CHCC criteria	Geographically matched healthy, unrelated subjects	109/234	NR	PR3-ANCA (79), MPO-ANCA (30)	1.07 (0.10 – 11.91)	
	Persson et al. 2013 ⁷	Case-Control	Sweden	ACR and CHCC criteria, EMA algorithm	Blood donors	105/200	GPA (61), MPA (44)	PR3-ANCA (62), MPO-ANCA (43)	0.63 (0.03 – 15.60)	
IL-6	Zhou et al. 2004 ⁶	Case-Control	USA	ACR criteria	Geographically and ethnically matched healthy, unrelated subjects	117/123	GPA (all patients)	PR3-ANCA (82), MPO-ANCA (5), PR3- and MPO-AN-CA positive (2), ANCA negative (25)	1.00 (0.70 – 1.44)	
rs1800795										
(C)										
	Spriewald et al. 2005 ³¹	Case-Control	Germany	ACR and CHCC criteria	Ethnically matched healthy subjects	32/91	GPA (all patients)	PR3-ANCA (all patients)	1.37 (0.76 – 2.48)	
	Previously unpublished Lyons et al. 2012 GWAS data ³	Case-control (GWAS)	UK	EMA algorithm, supported by either serology or histology	Wellcome Trust Case Control Consortium (UK)	914/5259	GPA (565), MPA (262)	PR3-ANCA (478), MPO-ANCA (264)	1.04 (0.94 – 1.15)	
	Husmann et al. 2014 ⁴⁰	Case-Control	Germany	EMA algorithm	Geographically matched healthy blood donors	863/1344	GPA (646), MPA (53), EGPA (164)	NR	0.95 (0.85 – 1.08)	
IL-10	Spriewald et al. 2005 ³¹	Case-Control	Germany	ACR and CHCC criteria	Ethnically matched healthy subjects	32/91	GPA (all patients)	PR3-ANCA (all patients)	1.10 (0.55 – 2.12)	
rs1800872										
(A)										
	Wiczorek et al. 2008 ⁴¹	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	506/507	GPA (403), EGPA (103)	ANCA+ (389), ANCA- (110)	1.00 (0.81 – 1.23)	

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Article	Study details		Definitions		Participants (n)		OR (95% CI)	
		Design	Setting	Cases	Controls	Cases / Controls	Diagnoses		ANCA serotype
IL-10 rs1800896 (G)	Murakozy et al. 2001 ⁴²	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	39/72	GPA (all patients)	NR	0.59 (0.34 – 1.03)
	Bartfai et al. 2003 ⁴³	Case-Control	Caucasian patients	ACR and CHCC criteria	Ethnically matched blood donors	161/153	GPA (125), MPA (36)	NR	0.74 (0.54 – 1.02)
	Spriewald et al. 2005 ³¹	Case-Control	Germany	ACR and CHCC criteria	Ethnically matched healthy subjects	32/91	GPA (all patients)	PR3-ANCA (all patients)	1.18 (0.67 – 2.10)
	Wieczorek et al. 2008 ⁴¹	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	506/507	GPA (403), EGPA (103)	ANCA+ (389), ANCA- (110)	0.88 (0.74 – 1.05)
	Previously unpublished Lyons et al. 2012 GWAS data ³	Case-control (GWAS)	UK	EMA algorithm, supported by either serology or histology	Wellcome Trust Case Control Consortium (UK)	676/5360	GPA (394), MPA (156)	PR3-ANCA (326), MPO-ANCA (167)	1.04 (0.93 – 1.17)
IRF5 rs10954213 (G)	Wieczorek et al. 2010 ⁴⁴	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	627/902	GPA (all patients)	NR	0.31 (0.26 – 0.37)
	Previously unpublished Lyons et al. 2012 GWAS data ³	Case-control (GWAS)	UK	EMA algorithm, supported by either serology or histology	Wellcome Trust Case Control Consortium (UK)	676/5365	GPA (394), MPA (156)	PR3-ANCA (326), MPO-ANCA (167)	1.04 (0.93 – 1.17)
	Kawasaki et al. 2013 ⁴⁵	Case-Control	Japan	EMA algorithm	Geographically matched healthy, unrelated subjects	232/710	GPA (28), MPA (177), EGPA (15), unclassified (12)	MPO (all patients)	1.28 (1.04 – 1.58)
	Wieczorek et al. 2010 ^{15***}	Case-Control	Germany	CHCC criteria	Geographically matched healthy subjects	456/818	GPA (all patients)	NR	0.72 (0.58 – 0.90)

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Article	Study details			Definitions		Cases / Controls		Participants (n)		OR (95% CI)
		Design	Setting	Cases	Controls	Cases / Controls	Diagnoses	ANCA serotype	Allele level		
<i>MPO</i> rs2333227 (A)	Reynolds et al. 2002 ⁴⁶	Case-Control	Germany	CHCC criteria	Geographically matched healthy subjects	226/508	GPA (all patients)	NR	0.71 (0.52 – 0.97)		
		Case-Control	Germany	ACR criteria	Geographically matched healthy subjects	196/1327	EGPA (all patients)	NR	1.41 (1.10 – 1.81)		
	Case-Control	The Netherlands	CHCC criteria	Ethnically matched healthy subjects	142/192	GPA (96), MPA (26), EGPA (8), iRPGN (12)	PR3-ANCA (92), MPO-ANCA (50)	1.07 (0.70 – 1.63)			
	Fiebel et al. 2004 ⁴⁷	Case-Control	Germany	Clinical criteria	Geographically matched healthy subjects	119/270	GPA (63), MPA (56)	PR3-ANCA (71), MPO-ANCA (48)	0.90 (0.62 – 1.32)		
<i>PCDI</i> rs1156882 (A)	Rajp et al. 2007 ⁴⁸	Case-Control	UK	CHCC criteria and histological criteria	Ethnically matched healthy subjects	134/150	GPA (69), MPA (65)	PR3-ANCA (91), MPO-ANCA (43)	0.92 (0.61 – 1.39)		
		Case-Control	The Netherlands	CHCC criteria	Geographically and ethnically matched healthy subjects	102/204	GPA (50), MPA (24), EGPA (7), RLV (21)	PR3-ANCA (49), MPO-ANCA (34), PR3- and MPO-AN-CA positive (2), ANCA negative (17)	0.94 (0.52 – 1.70)		
<i>PTPN22</i> rs2476601 (A)	Sakthivel et al. 2009 ⁴⁹	Case-Control	Sweden	Clinical criteria	Geographically and ethnically matched blood donors	66/275	GPA (all patients)	NR	0.78 (0.37 – 1.64)		
		Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy subjects	199/399	GPA (all patients)	c-ANCA+ (168), p-ANCA+ (1), ANCA negative (30)	1.75 (1.23 – 2.49)		
	Chung et al. 2012 ⁵⁰	Case-Control	USA	ACR criteria	Geographically matched subjects with no (family) history of autoimmune disease	424/469	GPA (all patients)	PR3-ANCA (339), MPO-ANCA (44), ANCA negative (39)	1.25 (0.91 – 1.71)		

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Article	Study details		Definitions		Cases / Controls		Participants (n)		OR (95% CI)	
		Design	Setting	Cases	Controls	Cases	Controls	Diagnoses	ANCA serotype	Allele level	
		Case-Control	Canada	ACR criteria	Geographically matched subjects with no history of autoimmune disease	456/1500	GPA (all patients)	NR		1.40 (1.11 – 1.78)	
	Previously unpublished Lyons et al. 2012 GWAS data ³	Case-control (GWAS)	UK	EMA algorithm, supported by either serology or histology	Wellcome Trust Case Control Consortium (UK)	676/5365	GPA (394), MPA (156)	PR3-ANCA (326), MPO-ANCA (167)		1.35 (1.14 – 1.60)	
	Martorana et al. 2012 ⁵¹	Case-Control	Italy	ACR and CHCC criteria	Geographically matched healthy subjects with no history of autoimmune disease	344/945	GPA (143), EGPA (99), MPA (102)	c-ANCA (126), p-ANCA (163), ANCA negative (39)		1.40 (0.97 – 2.04)	
RING1/	Heckmann et al. 2008 ²⁴	Case-Control	Germany	ACR criteria	Geographically matched healthy blood donors	279/369	GPA (all patients)	ANCA+ (250), ANCA- (27)		2.13 (1.70 – 2.67)	
RXRB											
rs213213 (A)	Previously unpublished Lyons et al. 2012 GWAS data ³	Case-control (GWAS)	UK	EMA algorithm, supported by either serology or histology	Wellcome Trust Case Control Consortium (UK)	676/5366	GPA (394), MPA (156)	PR3-ANCA (326), MPO-ANCA (167)		1.55 (1.38 – 1.74)	
	Xie et al. 2013 ²⁵	Case-control (GWAS)	Canada, USA	ACR criteria	Geographically matched healthy subjects	459/1503	GPA (all patients)	c-ANCA (351)		1.83 (1.57 – 2.13)	
RXRB	Szydl et al. 2006 ³²	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy subjects	187/201	GPA (all patients)	ANCA+ (151), ANCA- (36)		0.60 (0.45 – 0.80)	
rs6531 (C)	Previously unpublished Lyons et al. 2012 GWAS data ³	Case-control (GWAS)	UK	EMA algorithm, supported by either serology or histology	Wellcome Trust Case Control Consortium (UK)	911/5251	GPA (565), MPA (262)	PR3-ANCA (478), MPO-ANCA (264)		1.79 (1.61 – 1.98)	

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Article	Study details			Definitions		Participants (n)			OR (95% CI)
		Design	Setting	Cases	Controls	Cases / Controls	Diagnoses	ANCA serotype	Allele level	
RXRβ rs9277935 (T)	Xie et al. 2013 ²⁵	Case-control (GWAS)	Canada, USA	ACR criteria	Geographically matched healthy subjects	459/1503	GPA (all patients)	c-ANCA (351)	1.81 (1.55 – 2.11)	
	Wieczorek et al. 2009 ⁵³	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	282/380	GPA (all patients)	NR	0.39 (0.29 – 0.54)	
	Previously unpublished Lyons et al. 2012 GWAS data ³	Case-control (GWAS)	UK	EMA algorithm, supported by either serology or histology	Wellcome Trust Case Control Consortium (UK)	676/5350	GPA (394), MPA (156)	PR3-ANCA (326), MPO-ANCA (167)	0.51 (0.43 – 0.61)	
SERPINA1 S allele	Xie et al. 2013 ²⁵	Case-control (GWAS)	Canada, USA	ACR criteria	Geographically matched healthy subjects	459/1503	GPA (all patients)	c-ANCA (351)	0.34 (0.27 – 0.44)	
	Lhotta et al. 1994 ⁵⁴	Case-Control	Austria	ACR	Geographically matched healthy subjects	32/868	GPA (29), MPA (2), iRPGN (1)	c-ANCA (all patients)	0.69 (0.09 – 5.11)	
	Griffith et al. 1996 ⁵⁵	Case-Control	UK	CHCC criteria	Geographically and ethnically matched unrelated subjects	102/2310	GPA (51), MPA (29), EGPA (9), PAN (2), iRPGN (11)	c-ANCA (70), p-ANCA (32)	1.42 (0.79 – 2.52)	
	Mahr et al. 2010 ⁵⁶	Case-Control	USA	ACR criteria	Geographically matched healthy subjects with no (family) history of autoimmune disease	433/421	GPA (all patients)	PR3-ANCA (339), MPO-ANCA (44), ANCA negative (39)	1.68 (1.08 – 2.63)	
	Morris et al. 2011 ⁵⁷	Case-Control	Germany (531), France (81), UK (244)	CHCC criteria	Geographically and ethnically matched healthy subjects with no (family) history of autoimmune disease	856/1505	GPA (723), MPA (133)	c-ANCA (605), p-ANCA (150), ANCA negative (54)	1.11 (0.81 – 1.51)	

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Article		Study details		Definitions		Cases / Controls	Participants (n)		OR (95% CI)
	Design	Setting	Cases	Controls	Diagnoses	ANCA serotype		Allele level		
SERPINA1 Z allele	Chorostowska-Wynimko et al. 2013 ³⁸	Poland	Clinical, histological, and serological criteria	Geographically matched neonates born alive	GPA (all patients)	c-ANCA (43), p-ANCA (2), ANCA negative (4)	51/658		2.61 (0.56 – 12.08)	
	Lhotta et al. 1994 ⁵⁴	Austria	ACR criteria	Geographically matched healthy subjects	GPA (29), MPA (2), iRPGN (1)	c-ANCA (all patients)	32/868		8.76 (3.63 – 21.17)	
	Griffith et al. 1996 ⁵⁵	UK	CHCC criteria	Geographically and ethnically matched unrelated subjects	GPA (51), MPA (29), EGPA (9), PAN (2), iRPGN (11)	c-ANCA (70), p-ANCA (32)	102/2310		2.20 (1.05 – 4.62)	
	Callea et al. 1997 ⁵⁹	Italy	CHCC criteria	Healthy blood donors	GPA (33), MPA (28), iRPGN (23)	c-ANCA (38), p-ANCA (46)	84/200		4.90 (1.21 – 19.83)	
	Borgmann et al. 2001 ⁶⁰	Germany	ACR and CHCC criteria	Geographically and ethnically matched healthy, unrelated subjects	GPA (all patients)	c-ANCA (all patients)	97/752		3.36 (1.45 – 7.79)	
Mahr et al. 2010 ⁵⁶	USA	ACR criteria	Geographically matched healthy subjects with no (family) history of autoimmune disease	GPA (all patients)	PR3-ANCA (339), MPO-ANCA (44), ANCA negative (39)	433/421		1.99 (1.12 – 3.52)		
Morris et al. 2011 ⁵⁷	Germany (531), France (81), UK (244)	CHCC criteria	Geographically and ethnically matched healthy subjects with no (family) history of autoimmune disease	GPA (723), MPA (133)	c-ANCA (605), p-ANCA (150), ANCA negative (54)	856/1505		2.25 (1.60 – 3.18)		

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Article	Study details		Definitions		Cases / Controls	Participants (n)		OR (95% CI)
		Design	Setting	Cases	Controls		Diagnoses	ANCA serotype	
	Previously unpublished Lyons et al. 2012 GWAS data ^{3**}	Case-control (GWAS)	UK	EMA algorithm, supported by either serology or histology	Wellcome Trust Case Control Consortium (UK)	562/805	GPA (391), MPA (143)	PR3-ANCA (322), MPO-ANCA (166)	2.59 (1.66 – 4.02)
		Case-control (GWAS)	Europe	EMA algorithm, supported by either serology or histology	Geographically matched subjects	1445/1062	NR	NR	4.19 (2.62 – 6.69)
		Case-Control	Poland	Clinical, histological, serological criteria	Geographically matched neonates born alive	51/658	GPA (all patients)	c-ANCA (43), p-ANCA (2), ANCA negative (4)	2.19 (0.63 – 7.55)
STAT4 rs7574865 (T)	Wieczorek et al. 2010 ⁴⁴	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	612/880	GPA (all patients)	NR	1.01 (0.85 – 1.20)
	Previously unpublished Lyons et al. 2012 GWAS data ³	Case-control (GWAS)	UK	EMA algorithm, supported by either serology or histology	Wellcome Trust Case Control Consortium (UK)	676/5366	GPA (394), MPA (156)	PR3-ANCA (326), MPO-ANCA (167)	1.18 (1.04 – 1.35)
		Case-Control	Japan	EMA algorithm	Geographically matched healthy, unrelated subjects	232/710	GPA (28), MPA (177), EGPA (15), unclassified (12)	MPO (all patients)	1.10 (0.89 – 1.37)
		Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	39/72	GPA (all patients)	NR	2.20 (0.89 – 5.44)
TGF-β₁ rs1800471 (C)	Murakozy et al. 2001 ⁴²	Case-Control	Germany	ACR and CHCC criteria	Geographically matched healthy blood donors	161/96	GPA (125), MPA (36)	NR	1.06 (0.52 – 2.14)

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Article	Study details		Definitions		Participants (n)		OR (95% CI)	
		Design	Setting	Cases	Controls	Cases / Controls	Diagnoses	ANCA serotype	Allele level
TNFA rs1800629 (A)	Spriewald et al. 2005 ³¹	Case-Control	Germany	ACR and CHCC criteria	Ethnically matched healthy subjects	32/91	GPA (all patients)	PR3-ANCA (all patients)	1.86 (0.73 – 4.71)
	Mascher et al. 1997 ⁶¹	Case-Control	Germany	ACR and CHCC criteria	Healthy blood donors	35/111	GPA (all patients)	c-ANCA (32), p-ANCA (1), ANCA negative (1)	1.54 (0.77 – 3.10)
	Zhou et al. 2004 ⁶	Case-Control	USA	ACR criteria	Geographically and ethnically matched healthy, unrelated subjects	117/123	GPA (all patients)	PR3-ANCA (82), MPO-ANCA (5), PR3- and MPO-AN-CA positive (2), ANCA negative (25)	0.88 (0.52 – 1.50)
	Spriewald et al. 2005 ³¹	Case-Control	Germany	ACR and CHCC criteria	Ethnically matched healthy subjects	32/91	GPA (all patients)	PR3-ANCA (all patients)	2.22 (0.93 – 5.29)
TNFAII 196R	Previously unpublished Lyons et al. 2012 GWAS data ³	Case-control (GWAS)	UK	EMA algorithm, supported by either serology or histology	Wellcome Trust Case Control Consortium (UK)	676/5366	GPA (394), MPA (156)	PR3-ANCA (326), MPO-ANCA (167)	1.06 (0.92 – 1.23)
	Tsuchiya et al. 2003 ⁵	Case-Control	Japan	CHCC and Japanese 1998 criteria	Geographically matched healthy subjects	50/262	MPA (all patients)	MPO-ANCA (all patients)	0.67 (0.31 – 1.45)

Supplementary table 2. Study characteristics of the included studies (Continued)

Variants	Article	Study details			Definitions		Participants (n)			OR (95% CI)
		Design	Setting	Cases	Controls	Cases / Controls	Diagnoses	ANCA serotype	Allele level	
	Zhou et al. 2004 ⁶	Case-Control	USA	ACR criteria	Geographically and ethnically matched healthy, unrelated subjects	117/123	GPA (all patients)	PR3-ANCA (82), MPO-ANCA (5), PR3- and MPO-AN-CA positive (2), ANCA negative (25)	1.05 (0.70 – 1.57)	
<i>TLR9</i> rs352162 (T)	Husmann et al. 2014 ^{4,10**}	Case-Control	Germany	EMA algorithm	Geographically matched healthy blood donors	863/1344	GPA (646), MPA (53), EGPA (164)	NR	1.89 (1.67 – 2.14)	
<i>TLR9</i> rs352140 (T)	Husmann et al. 2014 ^{4,10**}	Case-Control	The Netherlands, UK, Germany	EMA algorithm	Geographically and ethnically matched healthy subjects	863/1344	GPA (273), MPA (100), EGPA (53)	NR	1.07 (0.90 – 1.28)	
<i>TLR9</i> rs352139 (T)	Husmann et al. 2014 ^{4,10**}	Case-Control	The Netherlands, UK, Germany	EMA algorithm	Geographically and ethnically matched healthy subjects	863/1344	GPA (646), MPA (53), EGPA (164)	NR	1.16 (1.03 – 1.31)	
<i>TLR9</i> rs5743836 (G)	Husmann et al. 2014 ^{4,10**}	Case-Control	The Netherlands, UK, Germany	EMA algorithm	Geographically and ethnically matched healthy subjects	863/1344	GPA (273), MPA (100), EGPA (53)	NR	1.08 (0.91 – 1.30)	
	Husmann et al. 2014 ^{4,10**}	Case-Control	Germany	EMA algorithm	Geographically matched healthy blood donors	863/1344	GPA (646), MPA (53), EGPA (164)	NR	0.82 (0.69 – 0.98)	
			The Netherlands, UK	EMA algorithm	Geographically and ethnically matched healthy subjects	426/554	GPA (273), MPA (100), EGPA (53)	NR	1.22 (0.94 – 1.58)	

ACR, American College of Rheumatology; ANCA, anti-neutrophil cytoplasm antibody; AAV, ANCA-associated vasculitis; c-ANCA, cytoplasmic ANCA; CHCC, Chapel Hill Consensus Conference; EGPA, eosinophilic granulomatosis with polyangiitis; EMA, European Medicines Agency; GPA, granulomatosis with polyangiitis; GWAS, genome-wide association study; IRPGN, idiopathic rapidly progressive glomerulonephritis; MPA, microscopic polyangiitis; MPO-ANCA, myeloperoxidase ANCA; NR, not reported; PAN, polyarteritis nodosa; p-ANCA, perinuclear ANCA; NR, not reported; RLV, renal limited vasculitis; PR3-ANCA, proteinase 3 ANCA; UK, United Kingdom; USA, United States of America. **Two cohorts described in the same publication; ***Three cohorts described in the same publication.

Supplementary table 3. Genetic variants not associated with AAV after meta-analysis

Variant by gene (minor allele)	Publications (n)	Cases (n) / Controls (n)	OR (95% CI)	P value for meta-analysis	I ² (%)	P value for heterogeneity	P value for funnel plot asymmetry
<i>CTLA-4</i> (AT) ₁₀₂	3	186/420	1.20 (0.75 – 1.90)	0.449	0	0.375	0.986
<i>CTLA-4</i> (AT) ₁₀₄	3	186/420	1.21 (0.92 – 1.60)	0.180	0	0.390	0.227
<i>CTLA-4</i> (AT) ₁₀₆	3	186/420	1.30 (0.74 – 2.29)	0.359	75	0.018	0.674
<i>CTLA-4</i> (AT) ₁₀₈	3	186/420	0.88 (0.50 – 1.56)	0.963	0	0.963	0.473
<i>CTLA-4</i> (AT) ₁₁₀	3	186/420	0.74 (0.34 – 1.61)	0.441	0	0.667	0.171
<i>CTLA-4</i> (AT) ₁₁₆	3	186/420	0.80 (0.27 – 2.33)	0.677	0	0.724	0.786
<i>CTLA-4</i> (AT) ₁₁₈	3	186/420	0.46 (0.16 – 1.35)	0.158	26	0.258	0.155
<i>CTLA-4</i> (AT) ₁₂₂	3	186/420	1.70 (0.62 – 4.71)	0.304	26	0.260	0.557
<i>CTLA-4</i> (AT) ₁₂₄	3	186/420	1.97 (0.73 – 5.31)	0.178	61	0.080	0.637
<i>CTLA-4</i> (AT) ₁₂₆	3	186/420	1.25 (0.53 – 2.94)	0.609	53	0.121	0.701
<i>CTLA-4</i> (AT) ₁₂₈	3	186/420	1.15 (0.47 – 2.82)	0.760	37	0.204	0.826
<i>FCAR</i> rs16986050 (G)	2	1311/5891	0.93 (0.82 – 1.05)	0.211	84	0.002	0.523
<i>FCGR3A</i> rs396991 (G)	2	141/457	1.00 (0.74 – 1.34)	0.978	79	0.030	N/A
<i>FCGR3B</i> (NA1)	4	982/970	0.96 (0.84 – 1.11)	0.590	37	0.191	0.210
<i>GHSR</i> rs509035 (A)	2	1547/6595	1.04 (0.96 – 1.14)	0.342	52	0.123	0.874
<i>GHSR</i> rs519384 (A)	1	628/1328	1.12 (0.96 – 1.30)	0.140	0	0.389	N/A
<i>GHSR</i> rs572169 (A)	1	620/1304	1.12 (0.97 – 1.30)	0.118	50	0.158	N/A
<i>HLA-A1</i>	4	427/7324	1.07 (0.89 – 1.30)	0.455	0	0.567	0.996
<i>HLA-A2</i>	5	443/7796	0.96 (0.82 – 1.12)	0.582	0	0.956	0.335
<i>HLA-A3</i>	4	427/7324	1.18 (0.97 – 1.42)	0.092	35	0.201	0.273
<i>HLA-A9</i>	3	366/6991	0.85 (0.65 – 1.13)	0.266	77	0.014	0.311
<i>HLA-A10</i>	3	366/6991	0.79 (0.48 – 1.30)	0.356	78	0.010	0.273

Supplementary table 3. Genetic variants not associated with AAV after meta-analysis (Continued)

Variant by gene (minor allele)	Publications (n)	Cases (n) / Controls (n)	OR (95% CI)	P value for meta-analysis	I ² (%)	P value for heterogeneity	P value for funnel plot asymmetry
HLA-A11	6	466/8107	0.97 (0.52 – 1.81)	0.924	65	0.013	0.273
HLA-A24	3	108/1223	0.94 (0.56 – 1.60)	0.830	0	0.510	0.090
HLA-A25	2	92/751	1.82 (0.72 – 4.58)	0.206	0	0.699	N/A
HLA-A26	4	131/1628	1.56 (0.89 – 2.74)	0.124	43	0.154	0.143
HLA-A28	3	376/6962	0.80 (0.52 – 1.23)	0.302	0	0.538	0.066
HLA-A29	3	123/988	0.88 (0.38 – 2.03)	0.760	42	0.178	0.801
HLA-A30	2	92/751	1.03 (0.25 – 4.28)	0.964	0	0.496	N/A
HLA-A31	3	108/1223	1.02 (0.43 – 2.44)	0.969	0	0.482	0.169
HLA-A32	3	108/1223	1.62 (0.74 – 3.54)	0.231	62	0.074	0.996
HLA-B7	4	427/7324	1.11 (0.91 – 1.35)	0.294	30	0.232	0.234
HLA-B12	2	335/6573	0.97 (0.76 – 1.23)	0.775	0	0.911	N/A
HLA-B13	4	427/7324	0.78 (0.46 – 1.34)	0.373	0	0.413	0.319
HLA-B14	4	427/7324	0.90 (0.54 – 1.49)	0.676	0	0.487	0.615
HLA-B15	2	355/5923	1.01 (0.77 – 1.31)	0.955	0	0.482	N/A
HLA-B18	4	427/7172	0.76 (0.50 – 1.16)	0.204	0	0.865	0.756
HLA-B27	4	412/7223	0.67 (0.44 – 1.02)	0.061	1	0.388	0.336
HLA-B35	4	412/7095	0.78 (0.58 – 1.06)	0.108	0	0.885	0.527
HLA-B37	2	355/5923	0.93 (0.51 – 1.70)	0.814	44	0.182	N/A
HLA-B39	2	67/523	0.65 (0.11 – 3.68)	0.622	0	0.619	N/A
HLA-B40	2	355/5923	0.94 (0.70 – 1.25)	0.652	0	0.319	N/A
HLA-B44	3	108/1223	1.53 (0.96 – 2.44)	0.072	46	0.157	0.225
HLA-B49	2	92/751	1.46 (0.43 – 4.92)	0.542	0	0.747	N/A

Supplementary table 3. Genetic variants not associated with AAV after meta-analysis (Continued)

Variant by gene (minor allele)	Publications (n)	Cases (n) / Controls (n)	OR (95% CI)	P value for meta-analysis	I ² (%)	P value for heterogeneity	P value for funnel plot asymmetry
HLA-B51	2	67/523	1.03 (0.46 – 2.32)	0.943	50	0.158	N/A
HLA-B55	2	67/523	1.07 (0.40 – 2.88)	0.887	93	<0.001	N/A
HLA-B57	2	92/751	2.37 (0.92 – 6.11)	0.075	0	0.493	N/A
HLA-B60	2	57/1172	1.98 (0.77 – 5.09)	0.154	20	0.263	N/A
HLA-B62	2	57/1172	1.95 (0.87 – 4.37)	0.107	0	0.688	N/A
HLA-Cw1	2	68/523	1.28 (0.59 – 2.77)	0.534	0	0.650	N/A
HLA-Cw3	2	68/523	1.36 (0.79 – 2.33)	0.267	0	0.907	N/A
HLA-Cw7	2	68/523	0.96 (0.57 – 1.62)	0.877	3	0.309	N/A
HLA-DPB1*0101	2	242/139	0.65 (0.34 – 1.22)	0.178	84	0.013	N/A
HLA-DPB1*0201	4	435/904	0.85 (0.65 – 1.12)	0.241	49	0.118	0.708
HLA-DPB1*0402	4	479/877	1.10 (0.87 – 1.40)	0.433	0	0.893	0.074
HLA-DPB1*0501	2	242/139	0.86 (0.22 – 3.33)	0.823	7	0.300	N/A
HLA-DPB1*0601	2	242/139	1.25 (0.28 – 5.67)	0.769	7	0.300	N/A
HLA-DPB1*0901	2	242/139	0.63 (0.20 – 2.01)	0.438	0	0.476	N/A
HLA-DQB1*02	2	83/141	0.91 (0.53 – 1.55)	0.720	0	0.796	N/A
HLA-DQB1*0302	2	126/141	1.03 (0.58 – 1.82)	0.931	0	0.832	N/A
HLA-DQB1*04	2	83/141	1.63 (0.52 – 5.11)	0.404	0	0.893	N/A
HLA-DQB1*0501	2	209/282	1.32 (0.79 – 2.20)	0.287	53	0.143	N/A
HLA-DQB1*0602	2	126/141	1.36 (0.70 – 2.64)	0.360	91	0.001	N/A
HLA-DQ ζ	3	149/546	0.82 (0.56 – 1.20)	0.302	49	0.142	0.252
HLA-DR1	10	1009/7105	0.93 (0.64 – 1.34)	0.684	70	<0.001	0.105
HLA-DR2	5	487/6222	1.10 (0.80 – 1.54)	0.554	37	0.176	0.757

Supplementary table 3. Genetic variants not associated with AAV after meta-analysis (Continued)

Variant by gene (minor allele)	Publications (n)	Cases (n) / Controls (n)	OR (95% CI)	P value for meta-analysis	I ² (%)	P value for heterogeneity	P value for funnel plot asymmetry
HLA-DR3	9	816/7045	1.05 (0.81 – 1.36)	0.740	41	0.096	0.946
HLA-DR4	11	914/7696	1.19 (0.95 – 1.50)	0.132	47	0.037	0.342
HLA-DR5	5	494/6155	0.90 (0.61 – 1.31)	0.570	32	0.206	0.965
HLA-DR7	7	610/6493	1.08 (0.73 – 1.60)	0.164	66	0.007	0.932
HLA-DR8	9	823/7157	1.10 (0.80 – 1.51)	0.552	15	0.307	0.375
HLA-DR9	11	1016/8204	1.16 (0.71 – 1.90)	0.560	60	0.005	0.925
HLA-DRB1*0401	2	216/465	0.93 (0.38 – 2.28)	0.869	48	0.167	N/A
HLA-DRB1*0403	2	216/465	0.49 (0.17 – 1.42)	0.190	53	0.144	N/A
HLA-DRB1*0405	2	216/465	0.78 (0.47 – 1.30)	0.344	27	0.242	N/A
HLA-DRB1*0406	2	216/465	1.22 (0.61 – 2.5)	0.580	44	0.182	N/A
HLA-DRB1*0407	2	216/465	1.95 (0.61 – 6.29)	0.263	13	0.283	N/A
HLA-DRB1*0410	2	216/465	1.17 (0.27 – 5.05)	0.832	0	0.463	N/A
HLA-DRB1*0802	2	216/465	0.94 (0.36 – 2.43)	0.897	0	0.359	N/A
HLA-DRB1*0803	2	216/465	1.33 (0.83 – 2.14)	0.236	0	0.622	N/A
HLA-DRB1*10	4	322/890	0.98 (0.39 – 2.46)	0.969	0	0.627	0.596
HLA-DRB1*11	4	322/890	0.98 (0.71 – 1.34)	0.818	0	0.818	1.000
HLA-DRB1*12	4	322/890	1.12 (0.60 – 2.09)	0.712	54	0.088	0.121
HLA-DRB1*1302	2	216/465	0.54 (0.27 – 1.05)	0.068	0	0.952	N/A
HLA-DRB1*1202	2	216/465	1.42 (0.81 – 2.49)	0.219	49	0.160	N/A
HLA-DRB1*1403	2	216/465	1.27 (0.43 – 3.76)	0.666	0	0.964	N/A
HLA-DRB1*1405	2	216/465	0.66 (0.32 – 1.37)	0.265	57	0.125	N/A
HLA-DRB1*1502	2	216/465	0.57 (0.30 – 1.07)	0.079	0	0.398	N/A

Supplementary table 3. Genetic variants not associated with AAV after meta-analysis (Continued)

Variant by gene (minor allele)	Publications (n)	Cases (n) / Controls (n)	OR (95% CI)	P value for meta-analysis	I ² (%)	P value for heterogeneity	P value for funnel plot asymmetry
<i>HLA-DRB1*16</i>	3	236/633	0.91 (0.37 – 2.27)	0.842	15	0.315	0.436
<i>HLA-DRB1*1602</i>	2	216/465	0.17 (0.02 – 1.29)	0.086	0	0.734	N/A
<i>HLA-DRB5</i>	2	110/1154	1.20 (0.73 – 1.97)	0.470	0	0.643	N/A
<i>IL-1β (A2)</i>	4	1172/5687	1.00 (0.90 – 1.12)	0.995	0	0.805	0.645
<i>ILIRN*1</i>	2	214/434	1.06 (0.82 – 1.37)	0.662	0	0.659	N/A
<i>ILIRN*2</i>	2	214/434	1.00 (0.77 – 1.31)	0.980	0	0.956	N/A
<i>ILIRN*3</i>	2	214/434	0.61 (0.26 – 1.44)	0.257	45	0.176	N/A
<i>ILIRN*4</i>	2	214/434	0.85 (0.13 – 5.82)	0.872	0	0.888	N/A
<i>ILIRN*5</i>	2	214/434	0.88 (0.13 – 5.94)	0.892	0	0.796	N/A
<i>IL-6 rs1800795 (C)</i>	4	1926/6817	1.01 (0.94 – 1.09)	0.793	0	0.537	0.584
<i>IL-10 rs1800872 (A)</i>	2	538/598	1.01 (0.83 – 1.22)	0.963	0	0.795	N/A
<i>IL-10 rs1800896 (G)</i>	5	1414/6183	0.90 (0.76 – 1.07)	0.232	55	0.063	0.285
<i>LEPR rs8179183 (C)</i>	1	878/2653	0.89 (0.77 – 1.03)	0.118	89	<0.001	0.780
<i>MPO rs2333227 (A)</i>	3	395/299	0.96 (0.76 – 1.21)	0.696	0	0.822	0.478
<i>PCD1 rs1156882 (A)</i>	2	168/479	0.88 (0.55 – 1.38)	0.568	0	0.701	N/A
<i>TGF-β_1 rs1800471 (C)</i>	3	232/259	1.49 (0.92 – 2.41)	0.107	0	0.401	0.220
<i>TNFA rs1800629 (A)</i>	4	860/5691	1.14 (0.88 – 1.45)	0.327	29	0.236	0.389
<i>TNFR1 196R</i>	2	167/385	0.94 (0.66 – 1.34)	0.749	0	0.317	N/A
<i>TLR9 rs5743836 (G)</i>	1	1289/1898	0.93 (0.81 – 1.08)	0.343	83	0.014	N/A

Supplementary table 4. Results of the meta-analyses stratified by diagnostic and serologic subgroups

Variant by gene	Publications (n)	Cases (n) / Controls (n)	OR (95% CI)	P value for meta-analysis
<i>CD226</i> rs763361 (T)				
GPA	3	2021/17898	1.19 (1.11 – 1.28)	<0.001
MPA	1	156/5366	1.04 (0.83 – 1.31)	0.739
EGPA	1	119/1226	1.19 (0.91 – 1.56)	0.208
PR3-ANCA	2	764/6592	1.22 (1.09 – 1.36)	<0.001
MPO-ANCA	2	199/6592	1.20 (0.99 – 1.46)	0.066
<i>CTLA-4</i> (AT) ₈₆				
GPA	3	210/432	0.44 (0.34 – 0.57)	<0.001
MPA	2	93/311	0.87 (0.60 – 1.25)	0.450
PR3-ANCA	1	62/200	0.71 (0.47 – 1.09)	0.122
MPO-ANCA	2	92/311	0.94 (0.65 – 1.35)	0.733
<i>CTLA-4</i> (AT) ₁₀₂				
GPA	2	93/309	0.94 (0.45 – 1.94)	0.856
MPA	2	93/311	0.86 (0.51 – 1.45)	0.574
PR3-ANCA	1	62/200	0.87 (0.41 – 1.87)	0.722
MPO-ANCA	2	92/311	0.69 (0.26 – 1.81)	0.456
<i>CTLA-4</i> (AT) ₁₀₄				
GPA	2	93/309	0.84 (0.55 – 1.27)	0.398
MPA	2	93/311	0.87 (0.58 – 1.29)	0.474
PR3-ANCA	1	62/200	0.42 (0.22 – 0.79)	0.007
MPO-ANCA	2	92/311	0.85 (0.57 – 1.26)	0.412
<i>CTLA-4</i> (AT) ₁₀₆				
GPA	2	93/309	1.69 (0.67 – 4.25)	0.264
MPA	2	93/311	0.64 (0.28 – 1.46)	0.287
PR3-ANCA	1	62/200	1.83 (0.60 – 5.55)	0.289
MPO-ANCA	2	92/311	0.53 (0.22 – 1.30)	0.168
<i>CTLA-4</i> (AT) ₁₀₈				
GPA	2	93/309	0.40 (0.12 – 1.35)	0.141
MPA	2	93/311	0.75 (0.36 – 1.54)	0.431
PR3-ANCA	1	62/200	0.30 (0.07 – 1.28)	0.103
MPO-ANCA	2	92/311	0.66 (0.31 – 1.41)	0.291
<i>CTLA-4</i> (AT) ₁₁₀				
GPA	2	93/309	0.55 (0.18 – 1.74)	0.311
MPA	2	93/311	0.26 (0.05 – 1.36)	0.109
PR3-ANCA	1	62/200	0.56 (0.16 – 1.94)	0.359
MPO-ANCA	2	92/311	0.26 (0.05 – 1.38)	0.113

Supplementary table 4. Results of the meta-analyses stratified by diagnostic and serologic subgroups (Continued)

Variant by gene	Publications (n)	Cases (n) / Controls (n)	OR (95% CI)	P value for meta-analysis
<i>CTLA-4</i> (AT) ₁₁₆				
GPA	2	93/309	0.66 (0.17 – 2.60)	0.553
MPA	2	93/311	0.41 (0.05 – 3.38)	0.408
PR3-ANCA	1	62/200	0.21 (0.01 – 3.72)	0.288
MPO-ANCA	2	92/311	0.69 (0.12 – 3.98)	0.673
<i>CTLA-4</i> (AT) ₁₁₈				
GPA	2	93/309	0.32 (0.06 – 1.68)	0.176
MPA	2	93/311	0.31 (0.06 – 1.69)	0.175
PR3-ANCA	1	62/200	0.11 (0.01 – 1.81)	0.121
MPO-ANCA	2	92/311	0.44 (0.10 – 1.97)	0.283
<i>CTLA-4</i> (AT) ₁₂₂				
GPA	2	93/309	1.32 (0.26 – 6.80)	0.742
MPA	2	93/311	1.57 (0.48 – 5.19)	0.456
PR3-ANCA	1	62/200	0.35 (0.02 – 6.62)	0.487
MPO-ANCA	2	92/311	3.69 (1.40 – 9.71)	0.008
<i>CTLA-4</i> (AT) ₁₂₄				
GPA	2	93/309	1.69 (0.67 – 4.25)	0.264
MPA	2	93/311	0.64 (0.28 – 1.46)	0.287
PR3-ANCA	1	62/200	1.83 (0.60 – 5.55)	0.289
MPO-ANCA	2	92/311	0.53 (0.22 – 1.30)	0.168
<i>CTLA-4</i> (AT) ₁₂₆				
GPA	2	93/309	1.43 (0.47 – 4.35)	0.526
MPA	2	93/311	0.38 (0.08 – 2.11)	0.266
PR3-ANCA	1	62/200	0.21 (0.01 – 3.72)	0.288
MPO-ANCA	2	92/311	0.54 (0.12 – 2.46)	0.428
<i>CTLA-4</i> (AT) ₁₂₈				
GPA	2	93/309	0.39 (0.07 – 2.14)	0.278
MPA	2	93/311	1.27 (0.39 – 4.20)	0.694
PR3-ANCA	1	62/200	0.29 (0.04 – 2.25)	0.235
MPO-ANCA	2	92/311	1.29 (0.39 – 4.28)	0.674
<i>CTLA-4</i> rs231775 (G)				
GPA	2	510/5994	1.05 (0.92 – 1.20)	0.430
MPA	2	252/5994	1.17 (0.97 – 1.40)	0.100
PR3-ANCA	1	326/5365	1.00 (0.85 – 1.18)	0.983
MPO-ANCA	1	167/5365	0.96 (0.77 – 1.21)	0.747

Supplementary table 4. Results of the meta-analyses stratified by diagnostic and serologic subgroups (Continued)

Variant by gene	Publications (n)	Cases (n) / Controls (n)	OR (95% CI)	P value for meta-analysis
<i>CTLA-4</i> rs3087243 (A)				
GPA	3	1561/7855	0.80 (0.73 – 0.87)	<0.001
MPA	2	358/5886	0.79 (0.68 – 0.92)	0.003
PR3-ANCA	1	478/5257	0.81 (0.71 – 0.93)	0.003
MPO-ANCA	1	264/5257	0.96 (0.80 – 1.15)	0.658
<i>FCAR</i> rs16986050 (G)				
GPA	2	1029/5891	0.92 (0.80 – 1.05)	0.214
MPA	1	394/5365	0.93 (0.69 – 1.26)	0.647
PR3-ANCA	1	326/5365	1.13 (0.93 – 1.38)	0.211
MPO-ANCA	1	167/5365	1.00 (0.75 – 1.33)	0.988
<i>FCGR2A</i> rs1801274 (C)				
GPA	5	1082/5969	0.99 (0.94 – 1.05)	0.827
MPA	4	800/5766	0.97 (0.81 – 1.15)	0.698
PR3-ANCA	4	883/5820	0.91 (0.79 – 1.03)	0.138
MPO-ANCA	4	844/5969	0.80 (0.67 – 0.95)	0.011
<i>FCGR3A</i> rs396991 (G)				
GPA	1	91/154	1.30 (0.89 – 1.91)	0.172
MPA	1	50/303	0.65 (0.39 – 1.07)	0.090
PR3-ANCA	1	91/154	1.30 (0.89 – 1.91)	0.172
MPO-ANCA	1	50/303	0.65 (0.39 – 1.07)	0.090
<i>FCGR3B</i> (NA1)				
GPA	3	865/667	0.93 (0.80 – 1.08)	0.343
MPA	2	75/200	1.52 (1.03 – 2.25)	0.035
PR3-ANCA	2	143/254	1.07 (0.79 – 1.45)	0.664
MPO-ANCA	2	121/403	1.19 (0.86 – 1.64)	0.287
<i>GHSR</i> rs509035 (A)				
GPA	2	1198/6595	1.05 (0.96 – 1.16)	0.292
MPA	1	262/5259	1.00 (0.82 – 1.20)	0.966
PR3-ANCA	1	478/5259	1.05 (0.91 – 1.21)	0.521
MPO-ANCA	1	264/5259	0.83 (0.68 – 1.01)	0.057
<i>HLA-A2</i>				
GPA	5	380/7796	0.96 (0.81 – 1.14)	0.633
c-ANCA	1	16/96	0.90 (0.37 – 2.23)	0.826
<i>HLA-A11</i>				
GPA	5	380/7702	0.61 (0.41 – 0.90)	0.014

Supplementary table 4. Results of the meta-analyses stratified by diagnostic and serologic subgroups (Continued)

Variant by gene	Publications (n)	Cases (n) / Controls (n)	OR (95% CI)	P value for meta-analysis
MPA	1	23/405	2.97 (1.19 – 7.41)	0.020
c-ANCA	1	16/472	0.71 (0.17 – 3.03)	0.644
<i>HLA-A24</i>				
GPA	3	108/1223	0.94 (0.56 – 1.60)	0.830
c-ANCA	1	16/472	0.75 (0.34 – 1.65)	0.476
<i>HLA-A26</i>				
GPA	3	108/1223	1.24 (0.61 – 2.54)	0.550
MPA	1	23/405	2.49 (1.00 – 6.18)	0.049
c-ANCA	1	16/472	2.36 (1.00 – 5.60)	0.051
<i>HLA-A31</i>				
GPA	3	108/1223	1.02 (0.43 – 2.44)	0.969
c-ANCA	1	16/472	1.55 (0.46 – 5.24)	0.479
<i>HLA-A32</i>				
GPA	3	108/1223	1.62 (0.74 – 3.54)	0.231
c-ANCA	1	16/472	2.07 (0.61 – 7.04)	0.246
<i>HLA-B35</i>				
GPA	4	349/7095	0.62 (0.44 – 0.89)	0.010
c-ANCA	1	16/472	0.80 (0.19 – 3.40)	0.757
<i>HLA-B39</i>				
GPA	2	67/523	0.65 (0.11 – 3.68)	0.622
c-ANCA	1	16/472	0.86 (0.11 – 6.51)	0.887
<i>HLA-B44</i>				
GPA	3	108/1223	1.53 (0.96 – 2.44)	0.072
c-ANCA	1	16/472	2.50 (0.85 – 7.40)	0.098
<i>HLA-B51</i>				
GPA	2	67/523	1.03 (0.46 – 2.32)	0.943
c-ANCA	1	16/472	1.87 (0.64 – 5.49)	0.255
<i>HLA-B55</i>				
GPA	2	67/523	1.07 (0.40 – 2.88)	0.887
c-ANCA	1	16/472	7.35 (2.33 – 23.14)	0.001
<i>HLA-B60</i>				
GPA	2	57/1172	1.98 (0.77 – 5.09)	0.154
c-ANCA	1	16/472	1.19 (0.28 – 5.13)	0.814
<i>HLA-B62</i>				
GPA	2	57/1172	2.27 (1.05 – 4.90)	0.036

Supplementary table 4. Results of the meta-analyses stratified by diagnostic and serologic subgroups (Continued)

Variant by gene	Publications (n)	Cases (n) / Controls (n)	OR (95% CI)	P value for meta-analysis
c-ANCA	1	16/472	2.21 (0.83 – 5.91)	0.114
<i>HLA-Cw1</i>				
GPA	2	68/523	1.28 (0.59 – 2.77)	0.534
c-ANCA	1	16/472	1.45 (0.58 – 3.58)	0.426
<i>HLA-Cw3</i>				
GPA	2	68/523	1.36 (0.79 – 2.33)	0.267
c-ANCA	1	16/472	1.40 (0.65 – 3.01)	0.384
<i>HLA-Cw7</i>				
GPA	2	68/523	0.96 (0.57 – 1.62)	0.877
c-ANCA	1	16/472	0.50 (0.12 – 2.10)	0.340
<i>HLA-DPA1 rs9277341</i>				
(C)				
GPA	2	1032/2200	0.35 (0.30 – 0.41)	<0.001
c-ANCA	1	578/1820	0.27 (0.22 – 0.33)	<0.001
<i>HLA-DPB1*0101</i>				
GPA	1	148/89	0.17 (0.05 – 0.63)	0.008
PR3-ANCA	2	183/139	0.45 (0.20 – 0.99)	0.048
MPO-ANCA	1	22/50	1.01 (0.29 – 3.48)	0.986
<i>HLA-DPB1*0201</i>				
GPA	2	283/458	0.67 (0.45 – 0.99)	0.042
MPA	1	50/77	1.38 (0.75 – 2.52)	0.298
EGPA	1	102/369	0.93 (0.57 – 1.51)	0.759
PR3-ANCA	1	148/89	0.44 (0.21 – 0.91)	0.027
MPO-ANCA	1	50/77	1.38 (0.75 – 2.52)	0.298
<i>HLA-DPB1*0301</i>				
GPA	3	774/918	0.23 (0.16 – 0.32)	<0.001
MPA	1	119/460	0.67 (0.37 – 1.21)	0.186
EGPA	2	167/829	0.47 (0.29 – 0.75)	0.001
PR3-ANCA	2	183/139	0.19 (0.09 – 0.39)	<0.001
MPO-ANCA	1	22/50	1.71 (0.74 – 3.94)	0.210
ANCA negative	1	27/369	0.58 (0.21 – 1.65)	0.311
<i>HLA-DPB1*0401</i>				
GPA	3	774/918	2.89 (2.50 – 3.35)	<0.001
MPA	1	119/460	1.09 (0.80 – 1.49)	0.597
EGPA	2	167/829	1.08 (0.85 – 1.36)	0.548

Supplementary table 4. Results of the meta-analyses stratified by diagnostic and serologic subgroups (Continued)

Variant by gene	Publications (n)	Cases (n) / Controls (n)	OR (95% CI)	P value for meta-analysis
PR3-ANCA	2	183/139	3.93 (2.75 – 5.62)	<0.001
MPO-ANCA	1	22/50	1.43 (0.67 – 3.02)	0.351
ANCA negative	1	27/369	1.26 (0.73 – 2.20)	0.407
<i>HLA-DPB1*0402</i>				
GPA	2	283/458	1.13 (0.82 – 1.55)	0.439
EGPA	1	102/369	1.15 (0.75 – 1.75)	0.533
PR3-ANCA	2	183/139	1.11 (0.69 – 1.77)	0.674
MPO-ANCA	1	22/50	0.94 (0.31 – 2.85)	0.913
<i>HLA-DPB1*0501</i>				
GPA	1	148/89	0.20 (0.01 – 4.93)	0.324
PR3-ANCA	2	183/139	1.11 (0.26 – 4.76)	0.886
MPO-ANCA	1	22/50	1.14 (0.10 – 12.91)	0.916
<i>HLA-DPB1*0601</i>				
GPA	1	148/89	0.60 (0.08 – 4.29)	0.610
PR3-ANCA	2	183/139	1.12 (0.23 – 5.51)	0.887
MPO-ANCA	1	22/50	6.93 (0.28 – 173.53)	0.239
<i>HLA-DPB1*0901</i>				
GPA	1	148/89	1.81 (0.07 – 44.73)	0.716
PR3-ANCA	2	183/139	0.98 (0.27 – 3.62)	0.978
MPO-ANCA	1	22/50	0.91 (0.17 – 4.85)	0.907
<i>HLA-DPB2 rs3130215</i>				
(A)				
GPA	3	1135/7249	1.37 (0.88 – 2.13)	0.160
MPA	1	156/5366	1.33(1.06 – 1.66)	0.013
PR3-ANCA	1	326/5366	0.65 (0.55 – 0.77)	<0.001
MPO-ANCA	1	167/5366	1.27 (1.02 – 1.58)	0.032
<i>HLA-DQB1*02</i>				
GPA	2	83/141	0.91 (0.53 – 1.55)	0.720
PR3-ANCA	1	32/91	0.98 (0.45 – 2.14)	0.954
<i>HLA-DQB1*0302</i>				
GPA	1	32/91	0.94 (0.36 – 2.49)	0.905
PR3-ANCA	2	67/141	1.03 (0.54 – 1.98)	0.924
MPO-ANCA	1	22/50	1.06 (0.37 – 2.99)	0.917
<i>HLA-DQB1*0303</i>				
GPA	1	32/91	2.97 (0.72 – 12.23)	0.132

Supplementary table 4. Results of the meta-analyses stratified by diagnostic and serologic subgroups (Continued)

Variant by gene	Publications (n)	Cases (n) / Controls (n)	OR (95% CI)	P value for meta-analysis
MPA	1	50/77	2.11 (1.14 – 3.90)	0.018
PR3-ANCA	2	67/141	0.79 (0.31 – 2.05)	0.634
MPO-ANCA	2	72/127	1.91 (1.12 – 3.26)	0.017
<i>HLA-DQB1*04</i>				
GPA	2	83/141	1.63 (0.52 – 5.11)	0.404
PR3-ANCA	1	32/91	1.74 (0.40 – 7.50)	0.457
<i>HLA-DQB1*0501</i>				
GPA	1	32/91	0.77 (0.32 – 1.88)	0.568
PR3-ANCA	2	67/141	1.05 (0.58 – 1.91)	0.865
MPO-ANCA	1	22/50	2.05 (0.84 – 4.97)	0.113
<i>HLA-DQB1*0602</i>				
GPA	1	32/91	0.89 (0.38 – 2.05)	0.779
PR3-ANCA	2	67/141	0.80 (0.42 – 1.54)	0.504
MPO-ANCA	1	22/50	0.35 (0.08 – 1.63)	0.181
<i>HLA-DQ7</i>				
GPA	1	32/91	0.43 (0.16 – 1.16)	0.095
MPA	1	23/405	0.66 (0.34 – 1.30)	0.234
PR3-ANCA	2	67/141	0.98 (0.58 – 1.66)	0.940
MPO-ANCA	1	22/50	1.26 (0.57 – 2.77)	0.569
<i>HLA-DR1</i>				
GPA	6	413/6002	0.76 (0.58 – 1.00)	0.054
MPA	4	223/6020	1.12 (0.72 – 1.74)	0.612
EGPA	3	128/5924	0.82 (0.53 – 1.26)	0.360
PR3-ANCA	3	157/560	0.57 (0.38 – 0.86)	0.008
MPO-ANCA	3	133/734	0.78 (0.49 – 1.25)	0.297
<i>HLA-DR2</i>				
GPA	4	301/6132	1.36 (1.08 – 1.72)	0.010
EGPA	1	14/113	1.40 (0.45 – 4.39)	0.561
PR3-ANCA	1	35/90	1.34 (0.63 – 2.86)	0.442
MPO-ANCA	1	22/90	1.03 (0.39 – 2.69)	0.958
c-ANCA	1	16/472	0.33 (0.08 – 1.38)	0.128
<i>HLA-DR3</i>				
GPA	5	368/5802	1.24 (1.00 – 1.56)	0.056
MPA	2	38/518	0.45 (0.17 – 1.14)	0.092
EGPA	2	116/482	0.70 (0.43 – 1.13)	0.139

Supplementary table 4. Results of the meta-analyses stratified by diagnostic and serologic subgroups (Continued)

Variant by gene	Publications (n)	Cases (n) / Controls (n)	OR (95% CI)	P value for meta-analysis
PR3-ANCA	3	157/560	1.29 (0.90 – 1.86)	0.170
MPO-ANCA	2	69/469	0.85 (0.49 – 1.48)	0.574
<i>HLA-DR4</i>				
GPA	7	425/6424	1.32 (1.09 – 1.60)	0.004
MPA	2	66/5555	1.50 (0.98 – 2.30)	0.060
EGPA	4	176/6246	1.70 (1.23 – 2.34)	0.001
PR3-ANCA	3	157/560	0.98 (0.71 – 1.36)	0.917
MPO-ANCA	2	69/202	2.51 (1.46 – 4.32)	0.001
c-ANCA	1	16/472	0.72 (0.27 – 1.88)	0.498
<i>HLA-DR5</i>				
GPA	3	285/5660	0.76 (0.54 – 1.06)	0.102
MPA	1	23/405	1.59 (0.80 – 3.15)	0.182
EGPA	1	14/113	1.24 (0.34 – 4.46)	0.746
PR3-ANCA	1	35/90	0.85 (0.30 – 2.42)	0.756
MPO-ANCA	1	22/90	0.52 (0.12 – 2.38)	0.402
<i>HLA-DR6</i>				
GPA	4	301/6132	0.45 (0.33 – 0.62)	<0.001
MPA	1	30/5442	0.29 (0.09 – 0.92)	0.035
EGPA	2	26/5555	0.19 (0.04 – 0.95)	0.043
PR3-ANCA	1	35/90	0.28 (0.06 – 1.25)	0.096
MPO-ANCA	1	22/90	0.70 (0.20 – 2.51)	0.586
c-ANCA	1	16/472	1.63 (0.56 – 4.77)	0.371
<i>HLA-DR7</i>				
GPA	5	368/5802	0.92 (0.71 – 1.19)	0.508
MPA	1	15/113	0.70 (0.20 – 2.44)	0.575
EGPA	3	164/804	1.71 (1.24 – 2.37)	0.001
PR3-ANCA	1	32/91	1.00 (0.40 – 2.48)	0.991
<i>HLA-DR8</i>				
GPA	5	381/6206	0.89 (0.57 – 1.39)	0.610
MPA	1	51/5442	0.31 (0.04 – 2.21)	0.242
EGPA	2	114/5811	3.08 (1.75 – 5.41)	<0.001
PR3-ANCA	3	157/560	0.68 (0.33 – 1.42)	0.308
MPO-ANCA	2	69/469	1.70 (0.80 – 3.60)	0.170
c-ANCA	1	16/472	1.63 (0.66 – 4.05)	0.291
<i>HLA-DR9</i>				

Supplementary table 4. Results of the meta-analyses stratified by diagnostic and serologic subgroups (Continued)

Variant by gene	Publications (n)	Cases (n) / Controls (n)	OR (95% CI)	P value for meta-analysis
GPA	6	402/6369	0.90 (0.58 – 1.39)	0.623
MPA	3	218/578	1.14 (0.83 – 1.56)	0.423
EGPA	3	164/832	1.07 (0.33 – 3.42)	0.915
PR3-ANCA	2	122/470	0.47 (0.09 – 2.58)	0.383
MPO-ANCA	3	175/1116	1.83 (1.29 – 2.60)	0.001
c-ANCA	1	16/472	3.03 (1.40 – 6.56)	0.005
<i>HLA-DRB1*0401</i>				
GPA	1	45/200	0.49 (0.06 – 3.90)	0.499
MPA	2	157/465	1.42 (0.60 – 3.41)	0.426
MPO-ANCA	1	64/265	2.10 (0.52 – 8.50)	0.300
<i>HLA-DRB1*0403</i>				
GPA	1	45/200	0.26 (0.02 – 4.46)	0.349
MPA	2	157/465	0.48 (0.14 – 1.59)	0.227
MPO-ANCA	1	64/265	0.95 (0.27 – 3.40)	0.943
<i>HLA-DRB1*0405</i>				
GPA	1	45/200	0.15 (0.01 – 2.49)	0.184
MPA	2	157/465	0.95 (0.56 – 1.62)	0.862
MPO-ANCA	1	64/265	0.61 (0.30 – 1.22)	0.159
<i>HLA-DRB1*0406</i>				
GPA	1	45/200	2.28 (0.67 – 7.74)	0.187
MPA	2	157/465	1.31 (0.62 – 2.78)	0.483
MPO-ANCA	1	64/265	0.65 (0.19 – 2.22)	0.487
<i>HLA-DRB1*0407</i>				
GPA	1	45/200	1.47 (0.06 – 36.42)	0.813
MPA	2	157/465	1.95 (0.54 – 6.99)	0.306
MPO-ANCA	1	64/265	2.82 (0.78 – 10.14)	0.113
<i>HLA-DRB1*0410</i>				
GPA	1	45/200	1.47 (0.06 – 36.42)	0.813
MPA	2	157/465	1.53 (0.35 – 6.64)	0.569
MPO-ANCA	1	64/265	1.67 (0.32 – 8.69)	0.544
<i>HLA-DRB1*0802</i>				
GPA	1	45/200	0.88 (0.04 – 18.50)	0.935
MPA	2	157/465	0.99 (0.35 – 2.79)	0.984
MPO-ANCA	1	64/265	1.16 (0.42 – 3.18)	0.778
<i>HLA-DRB1*0803</i>				

Supplementary table 4. Results of the meta-analyses stratified by diagnostic and serologic subgroups (Continued)

Variant by gene	Publications (n)	Cases (n) / Controls (n)	OR (95% CI)	P value for meta-analysis
GPA	1	45/200	1.18 (0.43 – 3.25)	0.749
MPA	2	157/465	1.25 (0.73 – 2.13)	0.413
MPO-ANCA	1	64/265	1.51 (0.78 – 2.93)	0.226
<i>HLA-DRB1*10</i>				
GPA	2	83/142	2.61 (0.45 – 15.24)	0.286
EGPA	1	102/369	0.40 (0.02 – 7.44)	0.538
PR3-ANCA	2	122/470	0.59 (0.13 – 2.65)	0.487
MPO-ANCA	1	47/379	1.48 (0.32 – 6.76)	0.616
<i>HLA-DRB1*11</i>				
GPA	2	83/142	0.97 (0.49 – 1.94)	0.932
EGPA	1	102/369	0.88 (0.53 – 1.48)	0.632
PR3-ANCA	2	122/470	0.97 (0.58 – 1.62)	0.908
MPO-ANCA	1	47/379	1.10 (0.51 – 2.38)	0.805
<i>HLA-DRB1*1101</i>				
GPA	1	45/200	0.16 (0.02 – 1.21)	0.076
MPA	2	223/465	2.57 (1.56 – 4.23)	<0.001
MPO-ANCA	1	116/265	2.79 (0.84 – 9.23)	0.093
<i>HLA-DRB1*12</i>				
GPA	2	83/142	4.06 (1.27 – 12.99)	0.018
EGPA	1	102/369	0.57 (0.17 – 1.93)	0.362
PR3-ANCA	2	122/470	1.01 (0.36 – 2.81)	0.983
MPO-ANCA	1	47/379	1.16 (0.26 – 5.16)	0.850
<i>HLA-DRB1*1201</i>				
GPA	1	45/200	0.15 (0.01 – 2.49)	0.184
MPA	2	157/465	0.51 (0.21 – 1.26)	0.146
MPO-ANCA	1	64/265	0.21 (0.03 – 1.60)	0.132
<i>HLA-DRB1*1202</i>				
GPA	1	45/200	2.92 (1.37 – 6.23)	0.005
MPA	2	157/465	1.07 (0.55 – 2.09)	0.838
PR3-ANCA	1	19/200	1.63 (0.46 – 5.75)	0.449
MPO-ANCA	2	89/465	1.00 (0.38 – 2.61)	1.000
<i>HLA-DRB1*13</i>				
GPA	2	83/142	0.61 (0.34 – 1.12)	0.111
EGPA	2	150/691	0.40 (0.24 – 0.68)	0.001
PR3-ANCA	1	32/91	0.56 (0.20 – 1.53)	0.257

Supplementary table 4. Results of the meta-analyses stratified by diagnostic and serologic subgroups (Continued)

Variant by gene	Publications (n)	Cases (n) / Controls (n)	OR (95% CI)	P value for meta-analysis
<i>HLA-DRB1*1302</i>				
GPA	1	45/200	0.89 (0.19 – 4.12)	0.878
MPA	2	157/465	0.46 (0.20 – 1.02)	0.055
MPO-ANCA	1	64/265	0.54 (0.24 – 1.23)	0.142
<i>HLA-DRB1*14</i>				
GPA	2	83/142	1.26 (0.40 – 4.01)	0.698
EGPA	1	102/341	1.83 (0.72 – 4.65)	0.205
PR3-ANCA	2	122/470	2.00 (0.87 – 4.56)	0.101
MPO-ANCA	1	47/379	2.04 (0.43 – 9.74)	0.372
<i>HLA-DRB1*1403</i>				
GPA	1	45/200	2.24 (0.20 – 24.93)	0.513
MPA	2	157/465	0.81 (0.21 – 3.19)	0.767
MPO-ANCA	1	64/265	1.25 (0.34 – 4.60)	0.739
<i>HLA-DRB1*1405</i>				
GPA	1	45/200	1.51 (0.53 – 4.27)	0.437
MPA	2	157/465	0.45 (0.18 – 1.15)	0.096
MPO-ANCA	1	64/265	0.11 (0.01 – 1.91)	0.131
<i>HLA-DRB1*15</i>				
GPA	2	83/142	0.97 (0.57 – 1.65)	0.921
PR3-ANCA	2	131/582	2.82 (2.00 – 3.96)	<0.001
MPO-ANCA	1	51/491	0.80 (0.37 – 1.73)	0.566
<i>HLA-DRB1*1501</i>				
GPA	1	45/200	1.70 (0.98 – 2.94)	0.059
MPA	2	157/465	1.65 (1.14 – 2.38)	0.007
PR3-ANCA	1	19/200	0.98 (0.40 – 2.45)	0.973
MPO-ANCA	2	89/465	2.03 (1.26 – 3.29)	0.004
<i>HLA-DRB1*1502</i>				
GPA	1	45/200	0.80 (0.18 – 3.69)	0.779
MPA	2	157/465	0.60 (0.30 – 1.21)	0.154
MPO-ANCA	1	64/265	0.67 (0.32 – 1.39)	0.279
<i>HLA-DRB1*16</i>				
GPA	2	83/142	0.23 (0.03 – 1.85)	0.166
PR3-ANCA	2	131/582	1.00 (0.29 – 3.41)	0.996
MPO-ANCA	1	51/491	3.55 (1.02 – 12.38)	0.047
<i>HLA-DRB1*1602</i>				

Supplementary table 4. Results of the meta-analyses stratified by diagnostic and serologic subgroups (Continued)

Variant by gene	Publications (n)	Cases (n) / Controls (n)	OR (95% CI)	P value for meta-analysis
GPA	1	45/200	0.40 (0.02 – 7.25)	0.533
MPA	2	157/465	0.23 (0.03 – 1.72)	0.151
MPO-ANCA	1	64/265	0.24 (0.01 – 4.17)	0.327
<i>HLA-DRB3</i>				
GPA	1	51/51	1.41 (0.76 – 2.60)	0.276
EGPA	2	150/691	0.58 (0.45 – 0.76)	<0.001
<i>HLA-DRB4</i>				
GPA	1	51/51	0.89 (0.46 – 1.72)	0.738
EGPA	2	150/691	2.06 (1.57 – 2.69)	<0.001
<i>IL-1β (A2)</i>				
GPA	3	714/5491	1.07 (0.93 – 1.22)	0.336
MPA	1	262/5259	0.89 (0.72 – 1.10)	0.286
PR3-ANCA	2	557/5455	1.01 (0.87 – 1.17)	0.930
MPO-ANCA	2	294/5455	1.00 (0.82 – 1.21)	0.971
<i>ILIRN*1</i>				
GPA	1	61/200	0.88 (0.57 – 1.37)	0.573
MPA	1	105/200	1.21 (0.83 – 1.77)	0.325
PR3-ANCA	2	122/434	1.12 (0.82 – 1.54)	0.476
MPO-ANCA	2	92/434	0.98 (0.69 – 1.40)	0.916
<i>ILIRN*2</i>				
GPA	1	61/200	1.22 (0.78 – 1.92)	0.380
MPA	1	105/200	0.77 (0.52 – 1.16)	0.212
PR3-ANCA	2	122/434	0.95 (0.68 – 1.31)	0.748
MPO-ANCA	2	92/434	1.02 (0.71 – 1.47)	0.921
<i>ILIRN*3</i>				
GPA	1	61/200	0.72 (0.15 – 3.40)	0.682
MPA	1	105/200	1.50 (0.55 – 4.08)	0.429
PR3-ANCA	2	122/434	0.61 (0.21 – 1.76)	0.358
MPO-ANCA	2	92/434	0.71 (0.22 – 2.29)	0.571
<i>ILIRN*4</i>				
GPA	1	61/200	0.65 (0.03 – 13.64)	0.782
MPA	1	105/200	1.91 (0.27 – 13.68)	0.518
PR3-ANCA	2	122/434	0.95 (0.11 – 8.63)	0.964
MPO-ANCA	2	92/434	3.08 (0.58 – 16.42)	0.188
<i>ILIRN*5</i>				

Supplementary table 4. Results of the meta-analyses stratified by diagnostic and serologic subgroups (Continued)

Variant by gene	Publications (n)	Cases (n) / Controls (n)	OR (95% CI)	P value for meta-analysis
GPA	1	61/200	1.09 (0.04 – 26.86)	0.959
MPA	1	105/200	0.63 (0.03 – 15.60)	0.779
PR3-ANCA	2	122/434	0.87 (0.10 – 7.74)	0.901
MPO-ANCA	2	92/434	2.18 (0.32 – 14.79)	0.424
<i>IL-6</i> rs1800795 (C)				
GPA	4	1360/1558	0.99 (0.94 – 1.04)	0.750
MPA	2	315/6593	0.98 (0.84 – 1.16)	0.829
EGPA	1	426/6593	0.96 (0.76 – 1.21)	0.730
PR3-ANCA	3	NR/6694	0.98 (0.89 – 1.09)	0.751
MPO-ANCA	2	NR/6603	1.04 (0.90 – 1.20)	0.616
<i>IL-10</i> rs1800872 (A)				
GPA	2	435/598	1.03 (0.84 – 1.27)	0.762
EGPA	1	103/507	0.89 (0.62 – 1.27)	0.512
PR3-ANCA	1	32/91	1.10 (0.55 – 2.20)	0.793
<i>IL-10</i> rs1800896 (G)				
GPA	5	993/6183	0.95 (0.85 – 1.05)	0.293
MPA	2	192/5513	0.95 (0.78 – 1.17)	0.655
EGPA	1	103/507	0.68 (0.50 – 0.92)	0.012
PR3-ANCA	2	358/5451	1.05 (0.90 – 1.22)	0.527
MPO-ANCA	1	167/5360	0.96 (0.77 – 1.19)	0.693
<i>IRF5</i> rs10954213 (G)				
GPA	2	1021/6267	0.66 (0.59 – 0.74)	<0.001
MPA	2	333/6075	1.22 (1.03 – 1.44)	0.018
PR3-ANCA	1	326/5365	1.09 (0.93 – 1.28)	0.293
MPO-ANCA	2	399/6075	1.12 (0.96 – 1.31)	0.142
<i>LEPR</i> rs8179183 (C)				
GPA	1	682/1326	0.72 (0.60 – 0.86)	<0.001
EGPA	1	196/1327	1.41 (1.10 – 1.81)	0.007
<i>MPO</i> rs2333227 (A)				
GPA	1	69/150	0.70 (0.41 – 1.21)	0.204
MPA	1	65/150	1.18 (0.72 – 1.93)	0.523
PR3-ANCA	3	258/549	1.00 (0.77 – 1.31)	0.977
MPO-ANCA	3	141/549	0.94 (0.68 – 1.31)	0.719
<i>PTPN22</i> rs2476601 (A)				
GPA	4	1616/8678	1.43 (1.26 – 1.62)	<0.001

Supplementary table 4. Results of the meta-analyses stratified by diagnostic and serologic subgroups (Continued)

Variant by gene	Publications (n)	Cases (n) / Controls (n)	OR (95% CI)	P value for meta-analysis
MPA	2	258/6310	1.42 (1.06 – 1.89)	0.018
EGPA	1	99/945	0.52 (0.21 – 1.29)	0.158
PR3-ANCA	2	419/6310	1.42 (1.14 – 1.77)	0.002
MPO-ANCA	2	183/6310	1.47 (1.08 – 2.00)	0.014
<i>RING1/RXR</i> rs213213 (A)				
GPA	3	1132/7238	1.91 (1.73 – 2.10)	<0.001
MPA	1	156/5366	1.15 (0.90 – 1.45)	0.264
PR3-ANCA	1	326/5366	2.06 (1.75 – 2.41)	<0.001
MPO-ANCA	1	167/5366	1.05 (0.83 – 1.32)	0.687
<i>RXR</i> rs6531 (C)				
GPA	3	1211/6955	1.70 (1.55 – 1.86)	<0.001
MPA	1	262/5251	1.38 (1.15 – 1.66)	0.001
PR3-ANCA	1	478/5251	2.19 (1.92 – 2.51)	<0.001
MPO-ANCA	1	264/5251	1.21 (1.00 – 1.46)	0.046
ANCA negative	1	36/201	1.02 (0.59 – 1.74)	0.948
<i>RXR</i> rs9277935 (T)				
GPA	3	1135/7233	0.37 (0.31 – 0.43)	<0.001
MPA	1	156/5350	0.93 (0.70 – 1.24)	0.629
PR3-ANCA	1	326/5350	0.24 (0.17 – 0.33)	<0.001
MPO-ANCA	1	167/5350	1.18 (0.92 – 1.53)	0.193
<i>SERPINA1</i> S allele				
GPA	2	484/1079	1.72 (1.12 – 2.66)	0.014
c-ANCA	3	145/3836	1.92 (1.12 – 3.29)	0.017
<i>SERPINA1</i> Z allele				
GPA	4	972/2636	2.40 (1.73 – 3.33)	<0.001
MPA	1	143/805	1.60 (0.76 – 3.39)	0.218
PR3-ANCA	1	322/805	2.58 (1.57 – 4.25)	<0.001
MPO-ANCA	1	166/805	2.01 (1.04 – 3.87)	0.037
c-ANCA	5	280/4788	3.53 (2.28 – 5.49)	<0.001
p-ANCA	2	78/2510	3.13 (1.21 – 8.13)	0.019
<i>STAT4</i> rs7574865 (T)				
GPA	2	1288/6246	1.06 (0.94 – 1.20)	0.331
MPA	1	676/5366	1.12 (0.86 – 1.46)	0.392
PR3-ANCA	1	676/5366	1.06 (0.88 – 1.28)	0.547

Supplementary table 4. Results of the meta-analyses stratified by diagnostic and serologic subgroups (Continued)

Variant by gene	Publications (n)	Cases (n) / Controls (n)	OR (95% CI)	P value for meta-analysis
MPO-ANCA	2	908/6076	1.24 (1.05 – 1.46)	0.009
<i>TGF-β₁</i> rs1800471 (C)				
GPA	3	196/259	1.52 (0.93 – 2.48)	0.096
MPA	1	36/96	1.03 (0.35 – 2.99)	0.960
PR3-ANCA	1	32/91	1.86 (0.73 – 4.71)	0.193
<i>TNFα</i> rs1800629 (A)				
GPA	4	578/5691	1.15 (0.98 – 1.35)	0.098
MPA	1	156/5366	1.13 (0.95 – 1.35)	0.182
PR3-ANCA	2	357/5457	1.21 (0.93 – 1.57)	0.149
MPO-ANCA	1	167/5366	0.88 (0.66 – 1.17)	0.375
<i>TNFRII</i> 196R				
GPA	1	177/123	1.05 (0.70 – 1.57)	0.819
MPA	1	50/262	0.67 (0.31 – 1.45)	0.313
MPO-ANCA	1	50/262	0.67 (0.31 – 1.45)	0.313
<i>TLR9</i> rs352162 (T)				
GPA	1	919/1898	1.20 (1.07 – 1.34)	0.001
MPA	1	153/1898	0.74 (0.58 – 0.95)	0.020
EGPA	1	217/1898	1.28 (1.05 – 1.57)	0.015
PR3-ANCA	1	NR/NR	1.30 (1.14 – 1.47)	<0.001
MPO-ANCA	1	NR/NR	0.79 (0.65 – 0.97)	0.028
ANCA negative	1	NR/NR	1.22 (0.99 – 1.50)	0.060
<i>TLR9</i> rs352140 (T)				
GPA	1	919/1898	1.20 (1.07 – 1.35)	0.001
MPA	1	153/1898	0.71 (0.55 – 0.91)	0.006
EGPA	1	217/1898	1.17 (0.96 – 1.43)	0.129
PR3-ANCA	1	NR/NR	1.28 (1.12 – 1.45)	<0.001
MPO-ANCA	1	NR/NR	0.75 (0.62 – 0.91)	0.004
ANCA negative	1	NR/NR	1.19 (0.97 – 1.47)	0.097
<i>TLR9</i> rs352139 (T)				
GPA	1	919/1898	1.18 (1.06 – 1.32)	0.003
MPA	1	153/1898	0.68 (0.52 – 0.87)	0.003
EGPA	1	217/1898	1.21 (0.99 – 1.47)	0.057
PR3-ANCA	1	NR/NR	1.23 (1.09 – 1.40)	0.001
MPO-ANCA	1	NR/NR	0.78 (0.63 – 0.96)	0.017
ANCA negative	1	NR/NR	1.20 (0.98 – 1.47)	0.086

Supplementary table 4. Results of the meta-analyses stratified by diagnostic and serologic subgroups (Continued)

Variant by gene	Publications (n)	Cases (n) / Controls (n)	OR (95% CI)	P value for meta-analysis
<i>TLR9</i> rs5743836 (G)				
GPA	1	919/1898	0.83 (0.70 – 0.99)	0.037
MPA	1	153/1898	1.78 (1.32 – 2.39)	<0.001
EGPA	1	217/1898	0.90 (0.68 – 1.21)	0.499
PR3-ANCA	1	NR/NR	0.83 (0.70 – 1.00)	0.045
MPO-ANCA	1	NR/NR	1.20 (0.91 – 1.58)	0.207
ANCA negative	1	NR/NR	0.99 (0.74 – 1.34)	0.970

Supplementary table 5. Results of the meta-analyses stratified by ethnic subgroups

Variant by gene	Publications (n)	Cases (n) / Controls (n)	OR (95% CI)	P value for meta-analysis
<i>CTLA-4</i> (AT) ₈₆				
Asian	1	49/111	1.31 (0.76 – 2.25)	0.333
Caucasian	3	254/432	0.46 (0.36 – 0.58)	<0.001
<i>CTLA-4</i> (AT) ₁₀₂				
Asian	1	49/111	0.89 (0.47 – 1.68)	0.714
Caucasian	2	137/309	1.73 (0.87 – 3.44)	0.117
<i>CTLA-4</i> (AT) ₁₀₄				
Asian	1	49/111	1.15 (0.70 – 1.91)	0.578
Caucasian	2	137/309	1.24 (0.88 – 1.73)	0.215
<i>CTLA-4</i> (AT) ₁₀₆				
Asian	1	49/111	0.41 (0.14 – 1.22)	0.109
Caucasian	2	137/309	2.69 (1.28 – 5.63)	0.009
<i>CTLA-4</i> (AT) ₁₀₈				
Asian	1	49/111	0.82 (0.33 – 2.02)	0.670
Caucasian	2	137/309	0.92 (0.44 – 1.92)	0.831
<i>CTLA-4</i> (AT) ₁₁₀				
Asian	1	49/111	0.25 (0.01 – 4.62)	0.349
Caucasian	2	137/309	0.84 (0.37 – 1.91)	0.679
<i>CTLA-4</i> (AT) ₁₁₆				
Asian	1	49/111	0.75 (0.03 – 18.56)	0.860
Caucasian	2	137/309	0.80 (0.26 – 2.50)	0.705
<i>CTLA-4</i> (AT) ₁₁₈				
Asian	1	49/111	0.75 (0.08 – 7.33)	0.807
Caucasian	2	137/309	0.41 (0.12 – 1.39)	0.152

Supplementary table 5. Results of the meta-analyses stratified by ethnic subgroups (Continued)

Variant by gene	Publications (n)	Cases (n) / Controls (n)	OR (95% CI)	P value for meta-analysis
<i>CTLA-4 (AT)₁₂₂</i>				
Asian	1	49/111	2.32 (0.57 – 9.45)	0.241
Caucasian	2	137/309	1.22 (0.27 – 5.49)	0.799
<i>CTLA-4 (AT)₁₂₄</i>				
Asian	1	49/111	2.28 (0.14 – 36.80)	0.562
Caucasian	2	137/309	1.94 (0.67 – 5.58)	0.222
<i>CTLA-4 (AT)₁₂₆</i>				
Asian	1	49/111	0.45 (0.05 – 3.88)	0.466
Caucasian	2	137/309	1.65 (0.63 – 4.32)	0.305
<i>CTLA-4 (AT)₁₂₈</i>				
Asian	1	49/111	6.98 (0.72 – 67.95)	0.094
Caucasian	2	137/309	0.72 (0.24 – 2.15)	0.560
<i>FCGR2A rs1801274 (C)</i>				
Asian	1	50/303	0.68 (0.37 – 1.24)	0.208
Caucasian	5	1126/5969	0.91 (0.82 – 1.00)	0.043
<i>FCGR3A rs396991 (G)</i>				
Asian	1	50/303	0.65 (0.39 – 1.07)	0.090
Caucasian	1	91/154	1.30 (0.89 – 1.91)	0.172
<i>FCGR3B (NA1)</i>				
Asian	1	50/303	1.29 (0.82 – 2.03)	0.267
Caucasian	3	865/667	0.93 (0.80 – 1.08)	0.343
<i>HLA-A2</i>				
Asian	1	16/472	0.90 (0.37 – 2.23)	0.826
Caucasian	4	427/7324	0.96 (0.81 – 1.13)	0.595
<i>HLA-A11</i>				
Asian	1	16/472	0.71 (0.17 – 3.03)	0.644
Caucasian	5	450/7635	1.01 (0.50 – 2.05)	0.978
<i>HLA-A24</i>				
Asian	1	16/472	0.75 (0.34 – 1.65)	0.476
Caucasian	2	92/751	1.16 (0.57 – 2.38)	0.683
<i>HLA-A26</i>				
Asian	1	16/472	2.36 (1.00 – 5.60)	0.051
Caucasian	3	115/1156	1.21 (0.58 – 2.55)	0.612
<i>HLA-A31</i>				
Asian	1	16/472	1.55 (0.46 – 5.24)	0.479
Caucasian	2	92/751	0.73 (0.21 – 2.52)	0.613

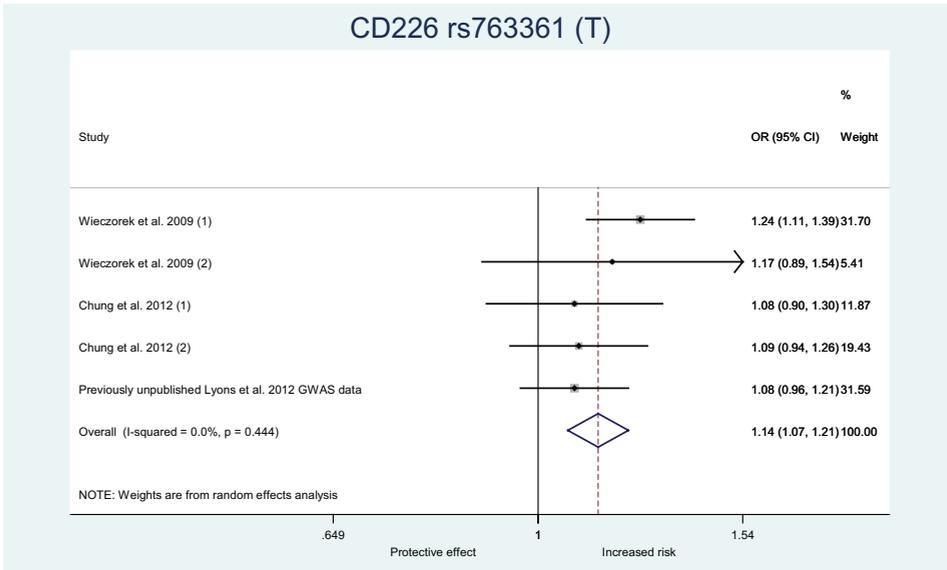
Supplementary table 5. Results of the meta-analyses stratified by ethnic subgroups (Continued)

Variant by gene	Publications (n)	Cases (n) / Controls (n)	OR (95% CI)	P value for meta-analysis
<i>HLA-A32</i>				
Asian	1	16/472	2.07 (0.61 – 7.04)	0.246
Caucasian	2	92/751	1.41 (0.52 – 3.84)	0.500
<i>HLA-B35</i>				
Asian	1	16/472	0.80 (0.17 – 3.40)	0.757
Caucasian	3	396/6623	0.78 (0.58 – 1.06)	0.115
<i>HLA-B39</i>				
Asian	1	16/472	0.86 (0.11 – 6.51)	0.887
Caucasian	1	51/51	0.33 (0.01 – 8.20)	0.499
<i>HLA-B44</i>				
Asian	1	16/472	2.50 (0.85 – 7.40)	0.098
Caucasian	2	92/751	1.41(0.85 – 2.35)	0.188
<i>HLA-B51</i>				
Asian	1	16/472	1.87 (0.64 – 5.49)	0.255
Caucasian	1	51/51	0.61 (0.19 – 1.92)	0.394
<i>HLA-B55</i>				
Asian	1	16/472	7.35 (2.34 – 23.14)	0.001
Caucasian	1	51/51	0.07 (0.00 – 1.30)	0.075
<i>HLA-B60</i>				
Asian	1	16/472	1.19 (0.28 – 5.13)	0.814
Caucasian	1	41/700	3.51 (0.99 – 12.36)	0.051
<i>HLA-B62</i>				
Asian	1	16/472	1.71 (0.58 – 4.99)	0.331
Caucasian	1	41/700	2.38 (0.70 – 8.12)	0.166
<i>HLA-Cw1</i>				
Asian	1	16/472	1.45 (0.58 – 3.58)	0.426
Caucasian	1	52/51	0.98 (0.24 – 4.03)	0.978
<i>HLA-Cw3</i>				
Asian	1	16/472	1.40 (0.66 – 3.01)	0.384
Caucasian	1	52/51	1.32 (0.62 – 2.81)	0.479
<i>HLA-Cw7</i>				
Asian	1	16/472	0.50 (0.12 – 2.10)	0.340
Caucasian	1	52/51	1.10 (0.62 – 1.96)	0.735
<i>HLA-DPBI*0201</i>				
Asian	1	50/77	1.38 (0.75 – 2.52)	0.298
Caucasian	3	385/827	0.76 (0.56 – 1.03)	0.073

Supplementary table 5. Results of the meta-analyses stratified by ethnic subgroups (Continued)

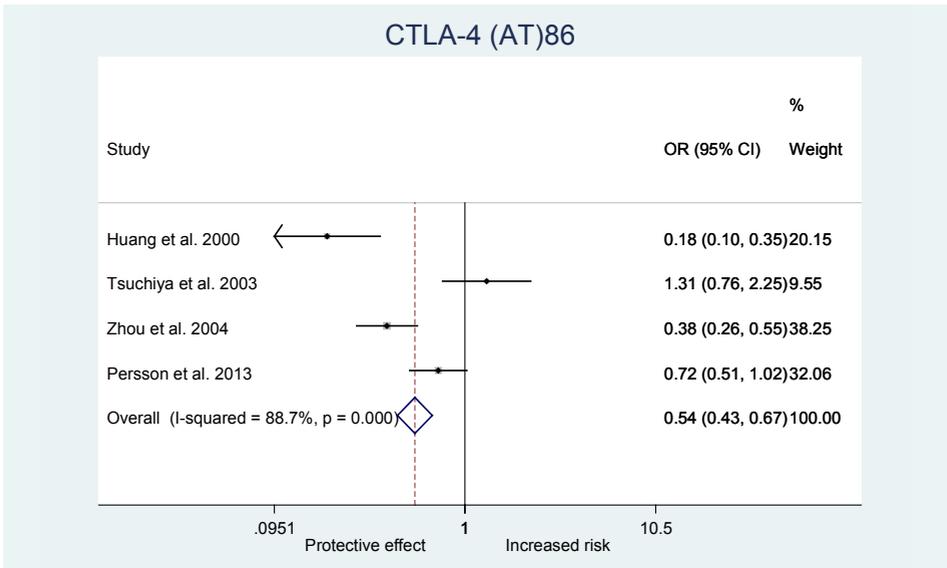
Variant by gene	Publications (n)	Cases (n) / Controls (n)	OR (95% CI)	P value for meta-analysis
<i>HLA-DQB1*0303</i>				
Asian	1	50/77	2.11 (1.14 – 3.90)	0.018
Caucasian	2	126/141	1.35 (0.55 – 3.33)	0.515
<i>HLA-DR1</i>				
Asian	1	64/265	1.12 (0.48 – 2.64)	0.793
Caucasian	9	945/6840	0.91 (0.61 – 1.36)	0.651
<i>HLA-DR2</i>				
Asian	1	16/472	0.33 (0.08 – 1.38)	0.128
Caucasian	4	471/5750	1.06 (0.87 – 1.29)	0.565
<i>HLA-DR4</i>				
Asian	1	16/472	0.72 (0.27 – 1.88)	0.498
Caucasian	10	882/7262	1.23 (0.97 – 1.57)	0.088
<i>HLA-DR6</i>				
Asian	1	16/472	1.63 (0.56 – 4.77)	0.371
Caucasian	4	471/5750	0.43 (0.30 – 0.61)	<0.001
<i>HLA-DR8</i>				
Asian	1	16/472	1.63 (0.66 – 4.05)	0.291
Caucasian	8	597/1483	1.05 (0.75 – 1.49)	0.768
<i>HLA-DR9</i>				
Asian	4	296/1409	1.67 (0.83 – 3.34)	0.148
Caucasian	7	510/1443	0.63 (0.34 – 1.19)	0.152
<i>IRF5 rs10954213 (G)</i>				
Asian	1	232/710	1.28 (1.04 – 1.58)	0.022
Caucasian	2	1303/6267	0.69 (0.63 – 0.76)	<0.001
<i>STAT4 rs7574865 (T)</i>				
Asian	1	232/710	1.10 (0.89 – 1.37)	0.388
Caucasian	2	1288/6246	1.11 (1.00 – 1.24)	0.044
<i>TNFR11 196R</i>				
Asian	1	50/262	0.67 (0.31 – 1.45)	0.313
Caucasian	1	117/123	1.05 (0.70 – 1.57)	0.819

Supplementary figure 1. Forest plots



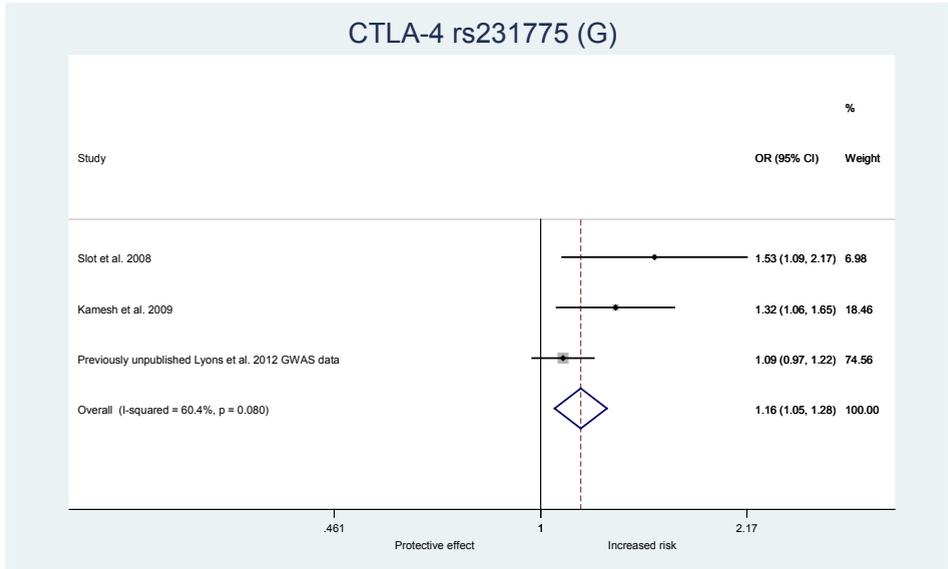
CD226 rs763361 (T) forest plot. Harbold test: N/A, Egger test: $p=0.792$.

References: ¹** , ²** , ³

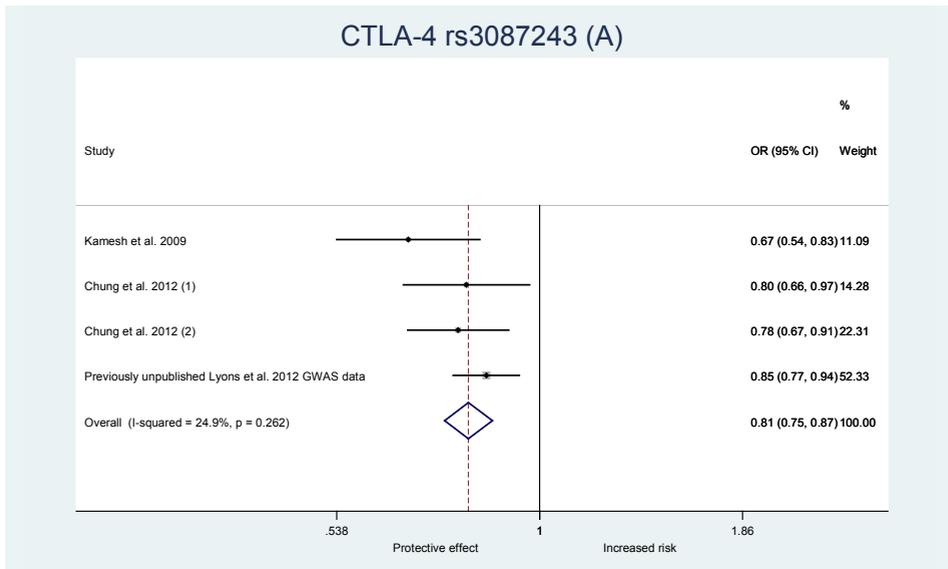


CTLA-4 (AT)₈₆ forest plot. Harbold test: $p=0.946$, Egger test: $p=0.788$.

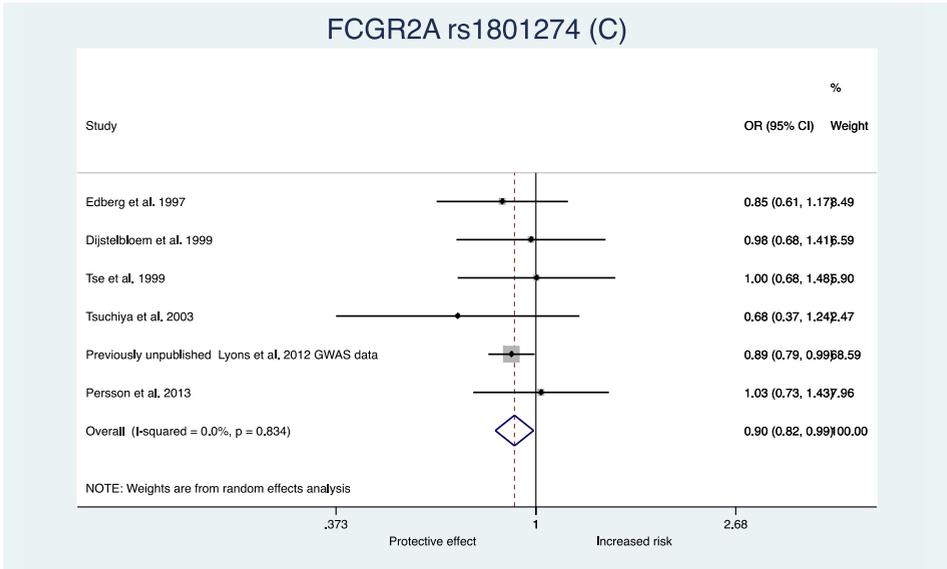
References: ⁴ , ⁵ , ⁶ , ⁷



CTLA-4 rs231775 (G) forest plot. Harbold test: $p=0.080$, Egger test: $p=0.081$.
References: ⁸, ⁹, ³

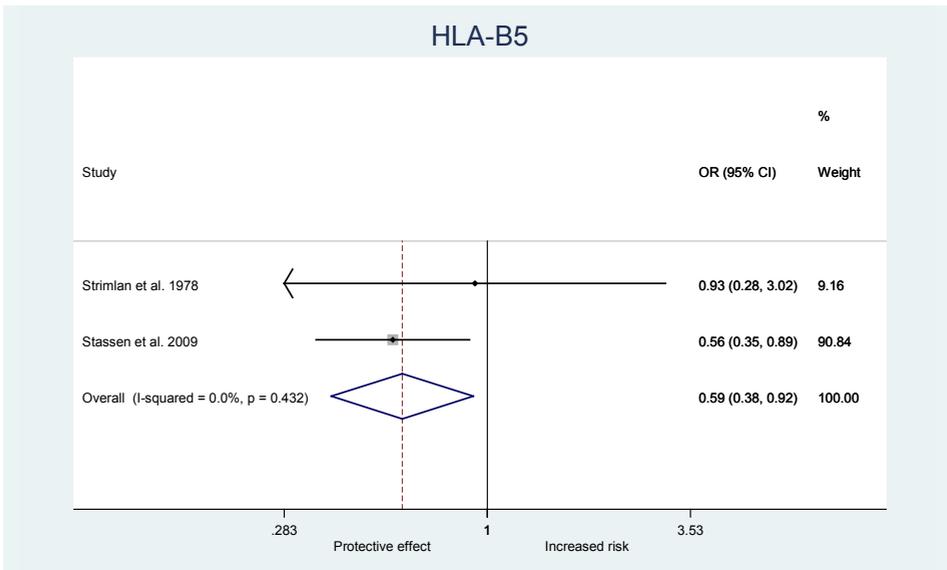


CTLA-4 rs3087243 (A) forest plot. Harbold test: N/A, Egger test: $p=0.122$.
References: ⁹, ^{2**}, ³



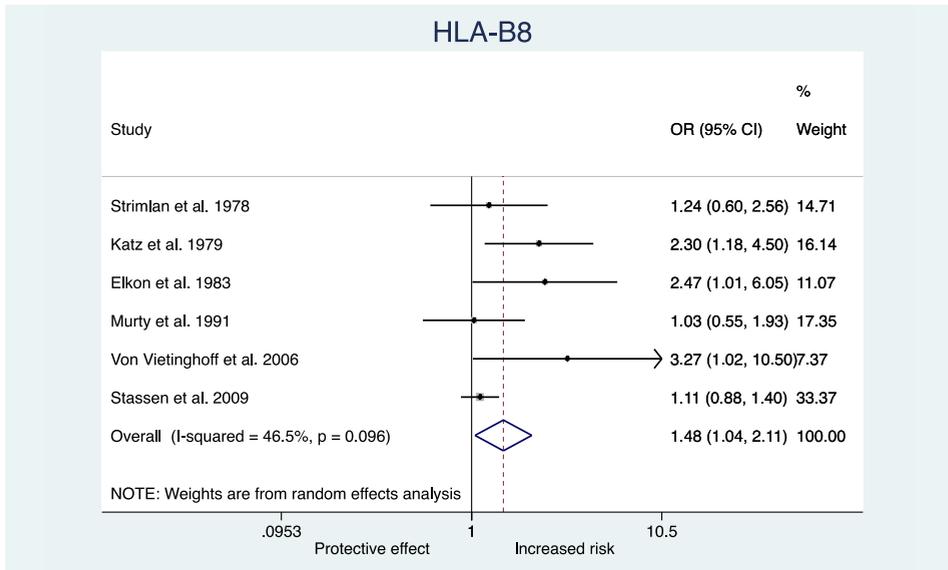
FCGR2A rs1801274 (C) forest plot. Harbord test: $p=0.788$, Egger test: $p=0.829$.

References: ^{11, 12, 13, 5, 7, 3}



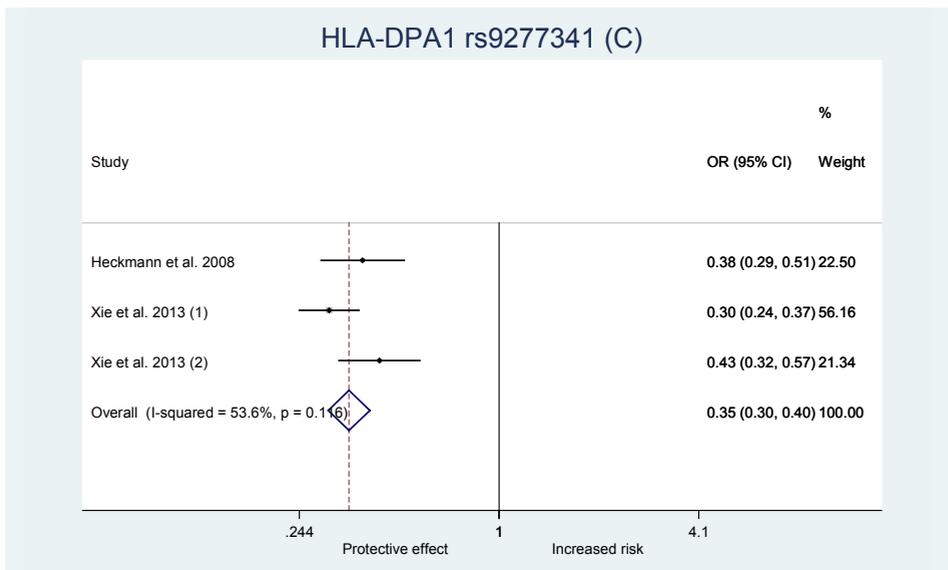
HLA-B5 forest plot. Harbord test: N/A, Egger test: N/A.

References: ^{16, 19}



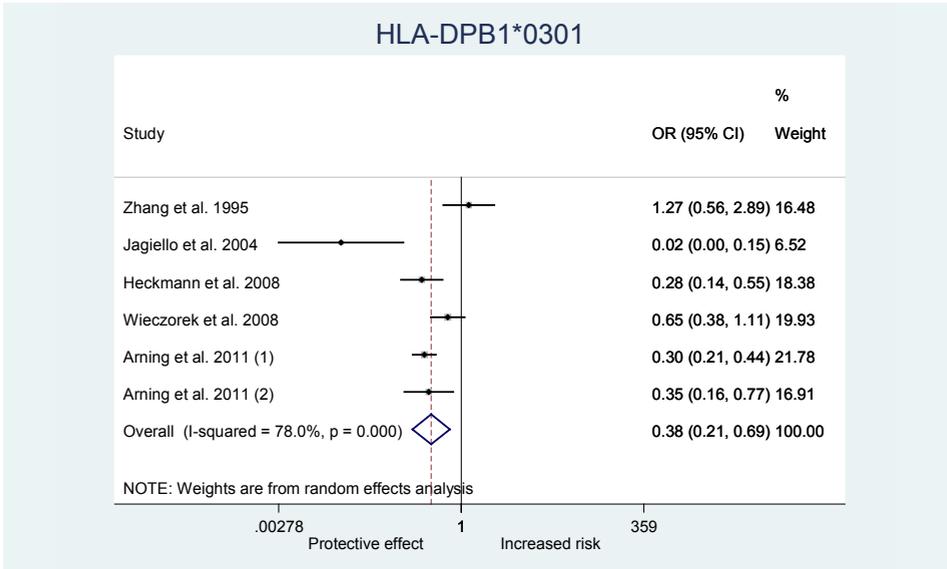
HLA-B8 forest plot. Harbold test: $p=0.063$, Egger test: $p=0.077$.

References: ^{16, 21, 23, 17, 18, 19}



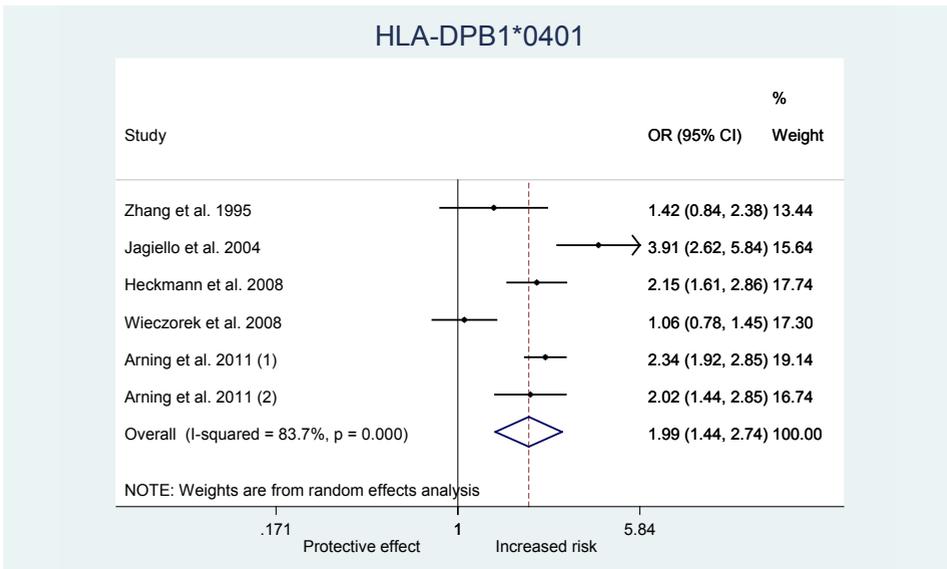
HLA-DPA1 rs9277341 (C) forest plot. Harbold test: $p=0.215$, Egger test: $p=0.219$.

References: ^{24, 25**}



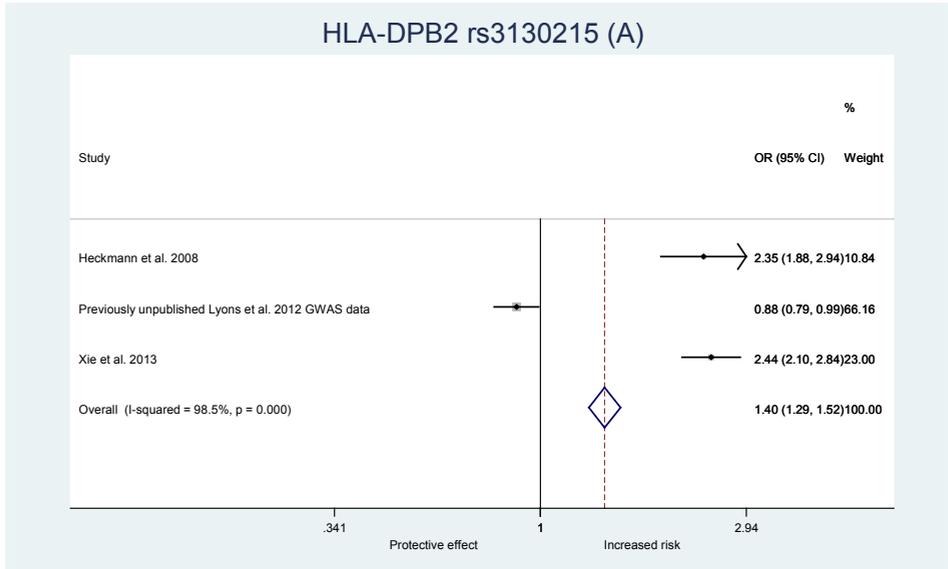
*HLA-DPB1*0301* forest plot. Harbold test: $p=0.938$, Egger test: $p=0.744$.

References: ²⁶, ²⁷, ²⁴, ²⁹, ^{30**}

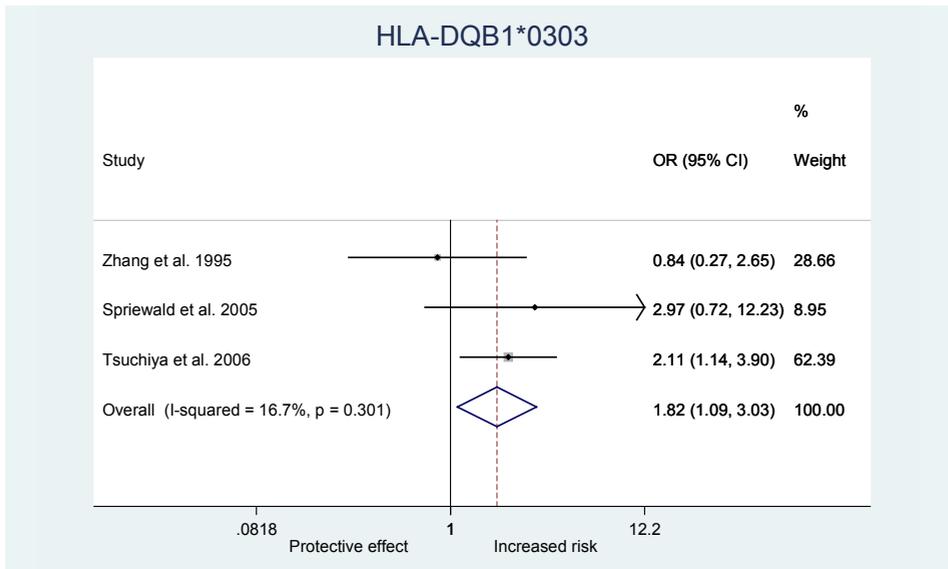


*HLA-DPB1*0401* forest plot. Harbold test: $p=0.738$, Egger test: $p=0.759$.

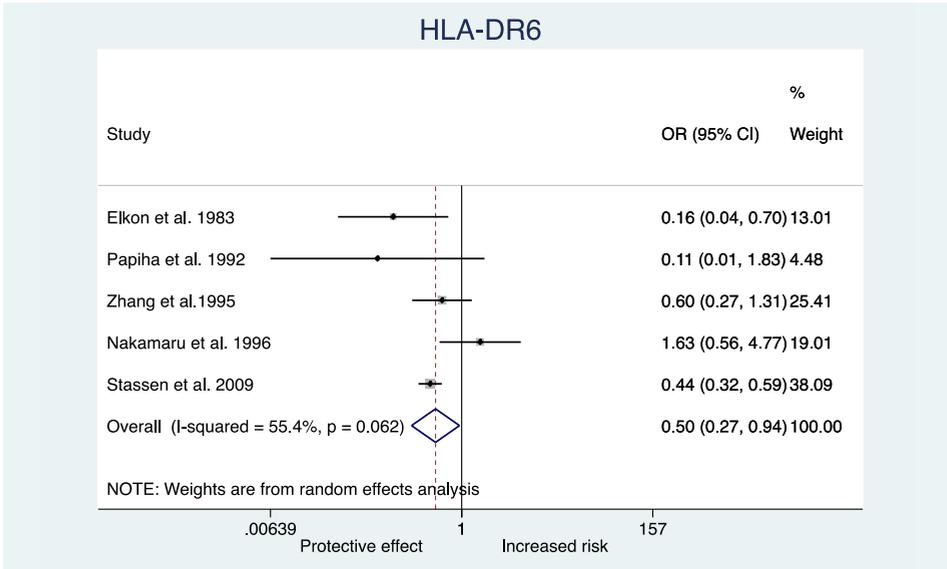
References: ²⁶, ²⁷, ²⁴, ²⁹, ^{30**}



HLA-DPB2 rs3130215 (A) forest plot. Harbold test: $p=0.446$, Egger test: $p=0.431$.
References: ^{24, 3, 25}

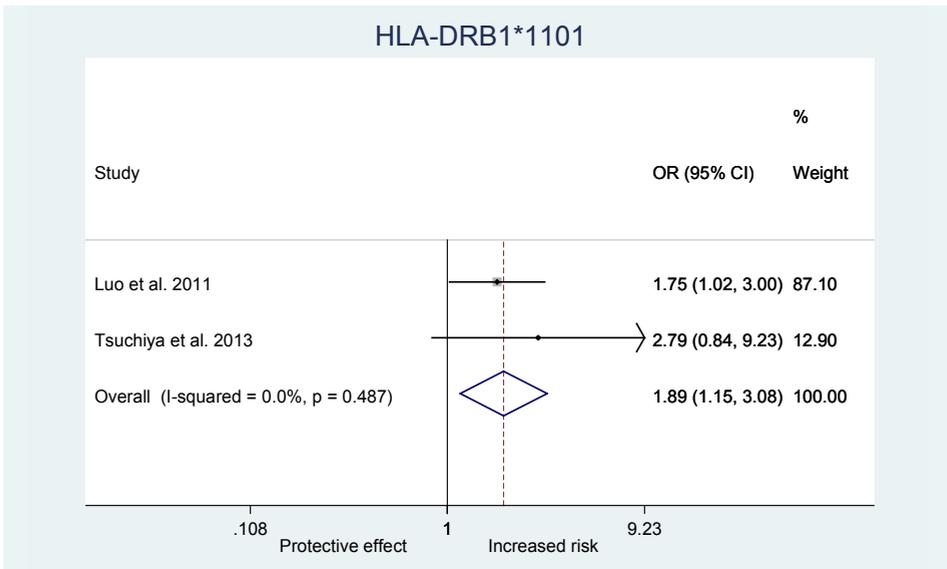


*HLA-DQB1*0303* forest plot. Harbold test: $p=0.916$, Egger test: $p=0.834$.
References: ^{26, 31, 28}



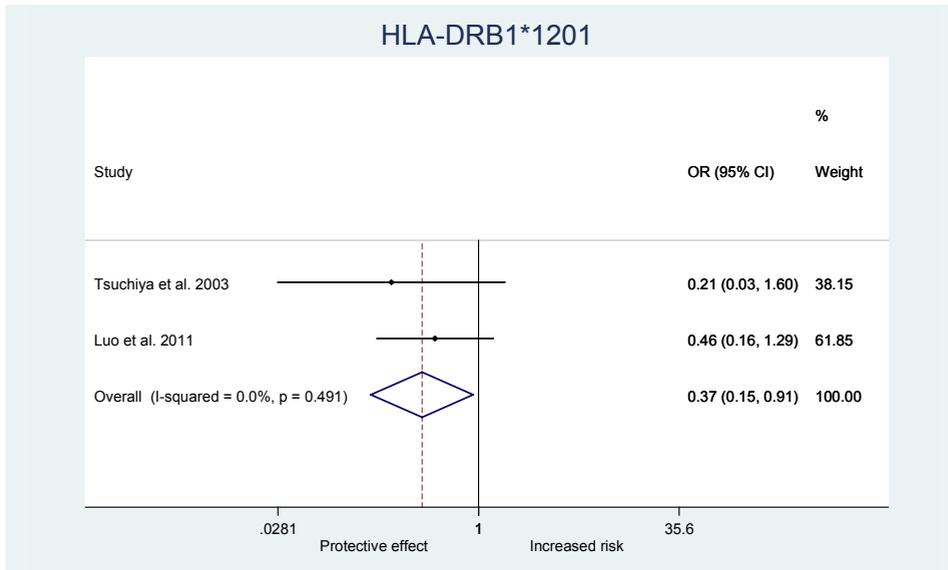
HLA-DR6 forest plot. Harbold test: $p=0.997$, Egger test: $p=0.989$.

References: ^{23, 32, 26, 20, 19}

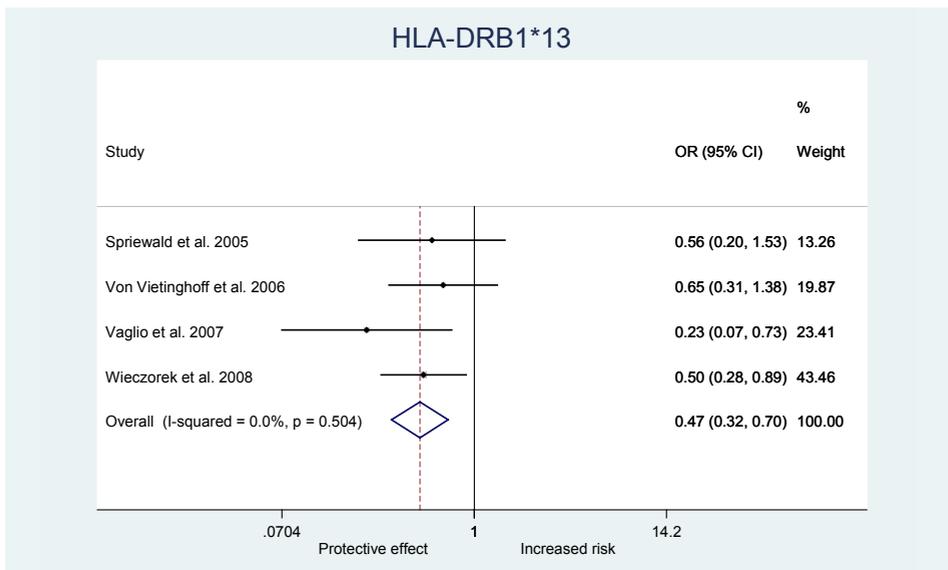


*HLA-DRB1*1101* forest plot. Harbold test: N/A, Egger test: N/A.

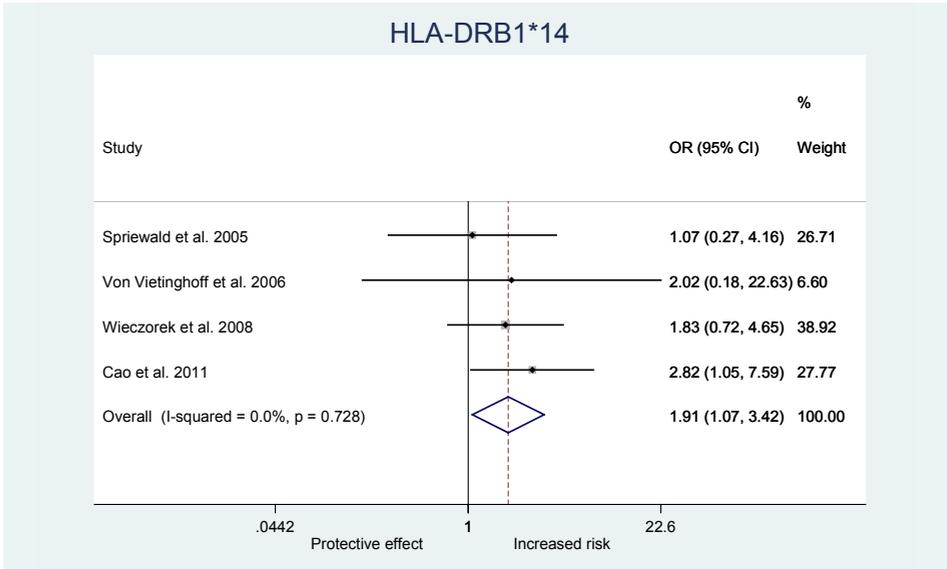
References: ^{34, 37}



*HLA-DRB1*1201* forest plot. Harbord test: N/A, Egger test: N/A.
References: ^{5, 34}

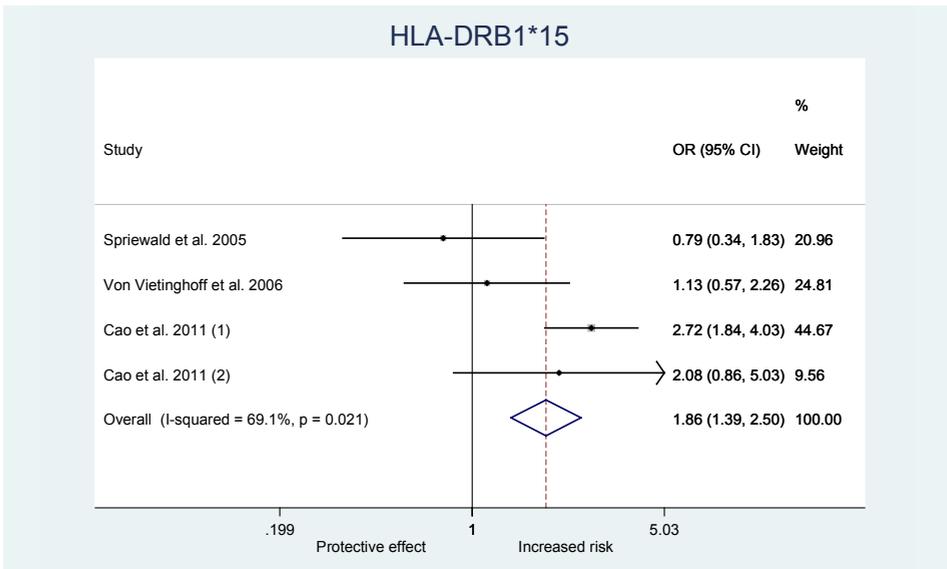


*HLA-DRB1*13* forest plot. Harbord test: $p=0.884$, Egger test: $p=0.467$.
References: ^{31, 18, 35, 29}



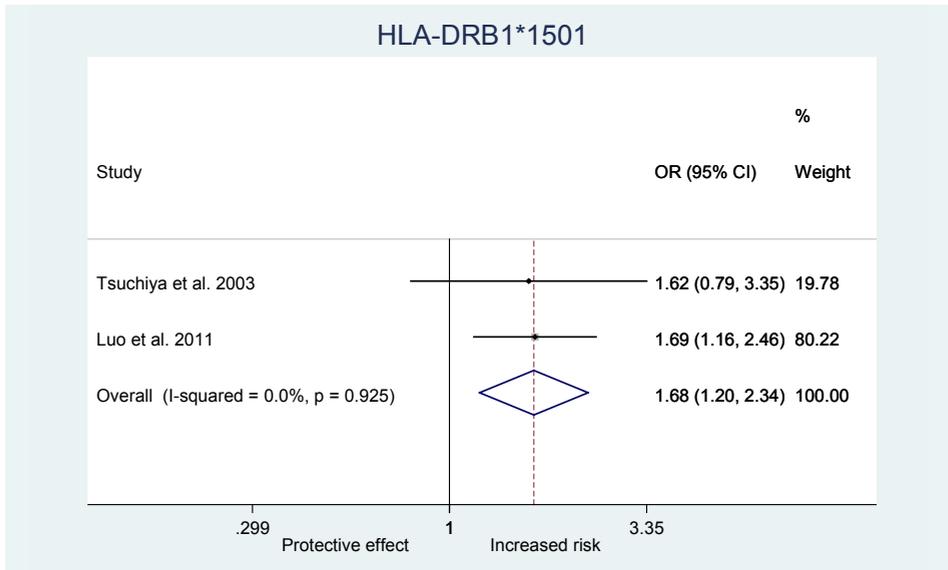
*HLA-DRB1*14* forest plot. Harbord test: $p=0.700$, Egger test: $p=0.706$.

References: ³¹, ¹⁸, ²⁹, ³³

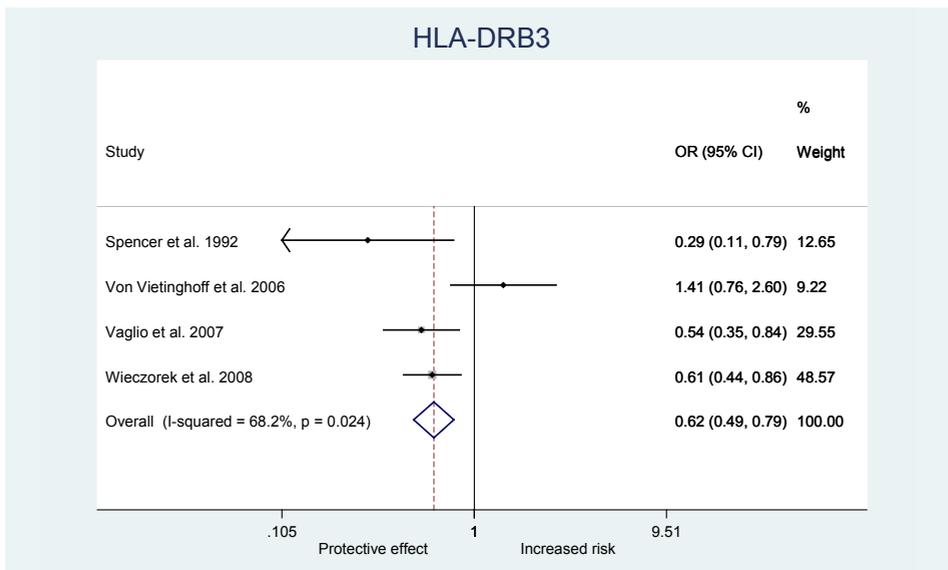


*HLA-DRB1*15* forest plot. Harbord test: $p=0.347$, Egger test: $p=0.214$.

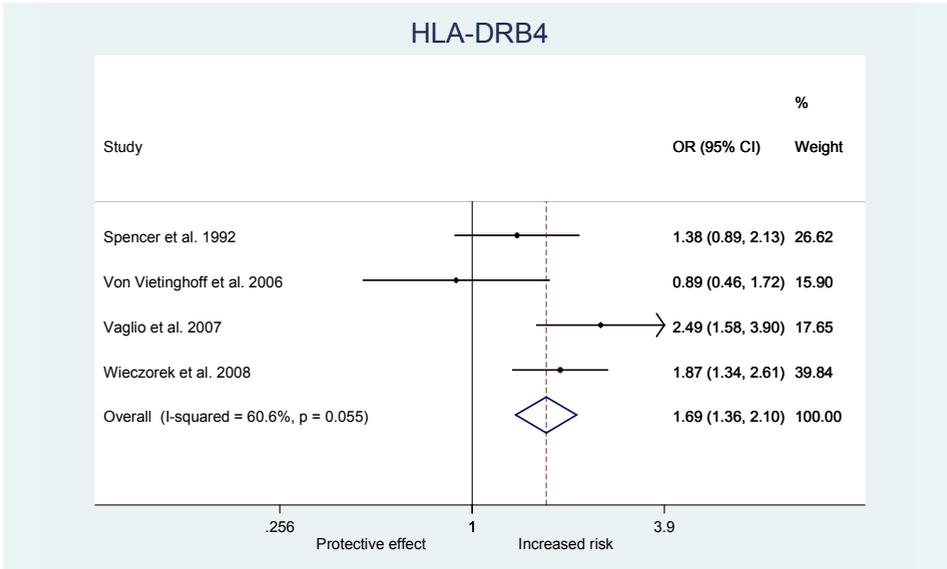
References: ³¹, ¹⁸, ^{33**}



*HLA-DRB1*1501* forest plot. Harbold test: N/A, Egger test: N/A.
References: ^{5, 34}

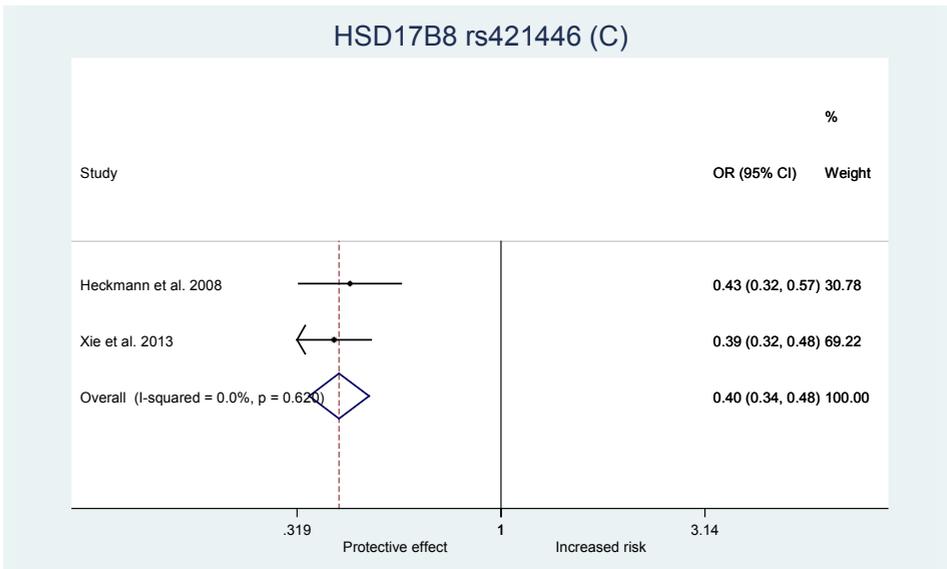


HLA-DRB3 forest plot. Harbold test: $p=0.689$, Egger test: $p=0.958$.
References: ^{38, 18, 35, 29}



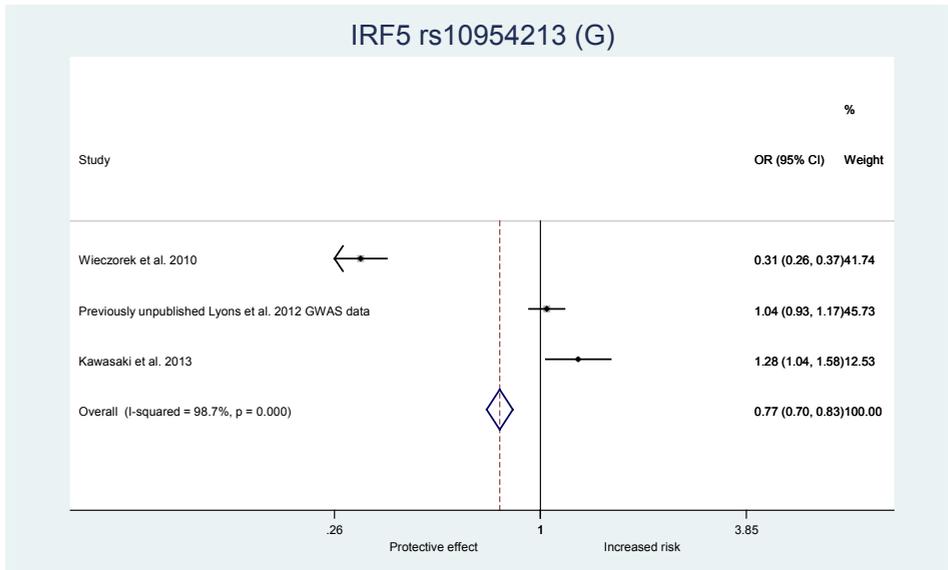
HLA-DRB4 forest plot. Harbold test: $p=0.533$, Egger test: $p=0.388$.

References: ^{38, 18, 35, 29}

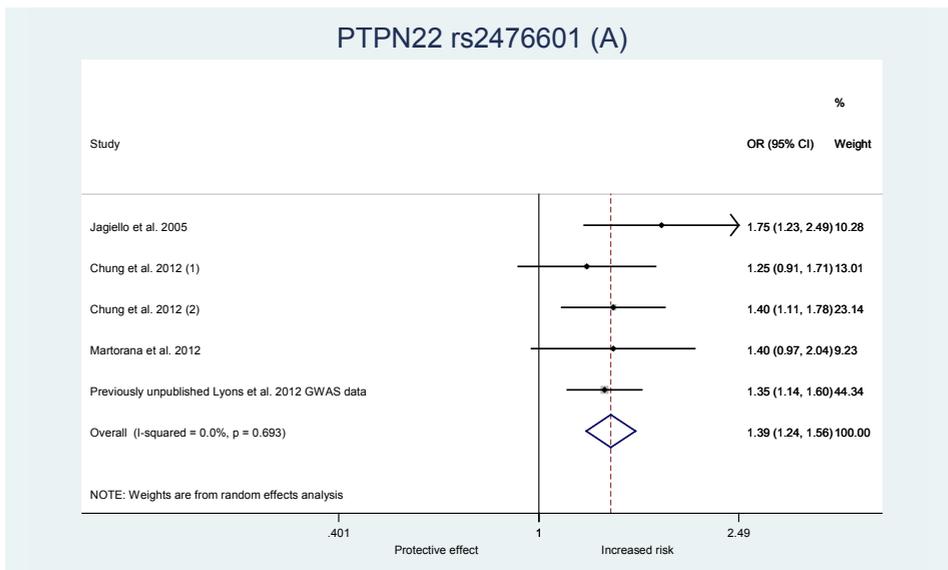


HSD17B8 rs421446 (C) forest plot. Harbold test: N/A, Egger test: N/A.

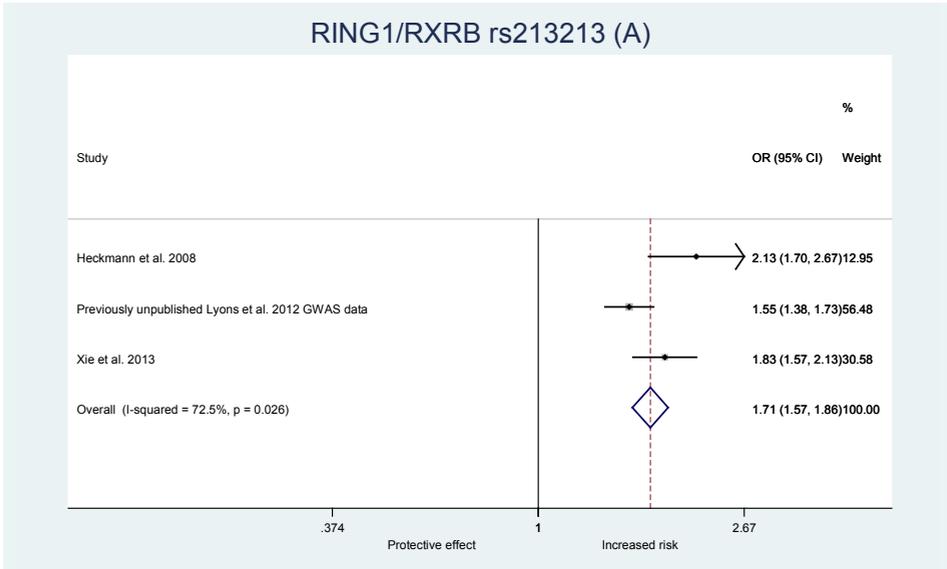
References: ^{24, 25}



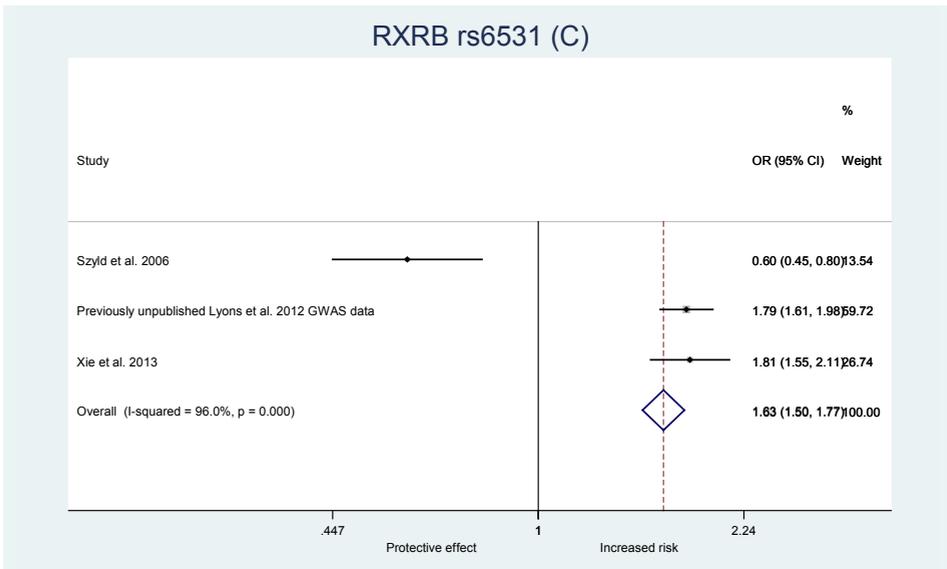
IRF5 rs10954213 (G) forest plot. Harbold test: $p=0.948$, Egger test: $p=0.833$.
References: ⁴⁴, ³, ⁴⁵



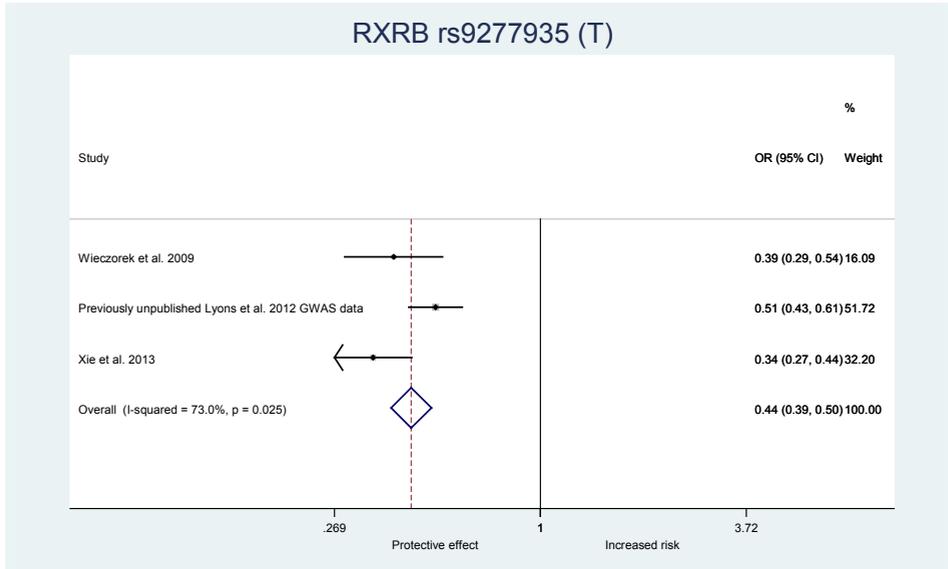
PTPN22 rs2476601 (A) forest plot. Harbold test: N/A, Egger test: $p=0.500$.
References: ⁵⁰, ^{2**}, ⁵¹, ³



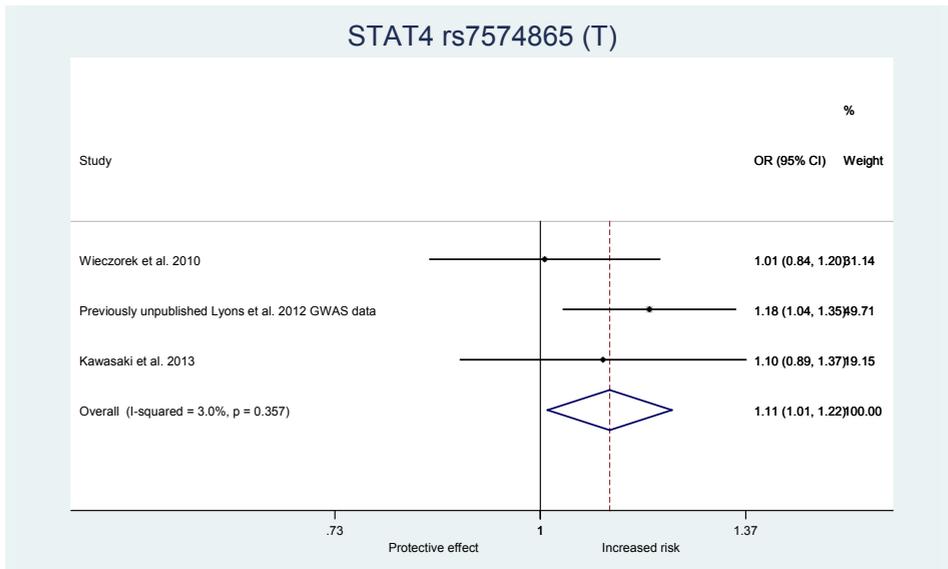
RING1/RXR_B rs213213 (A) forest plot. Harbold test: $p=0.187$, Egger test: $p=0.169$.
References: ²⁴, ³, ²⁵



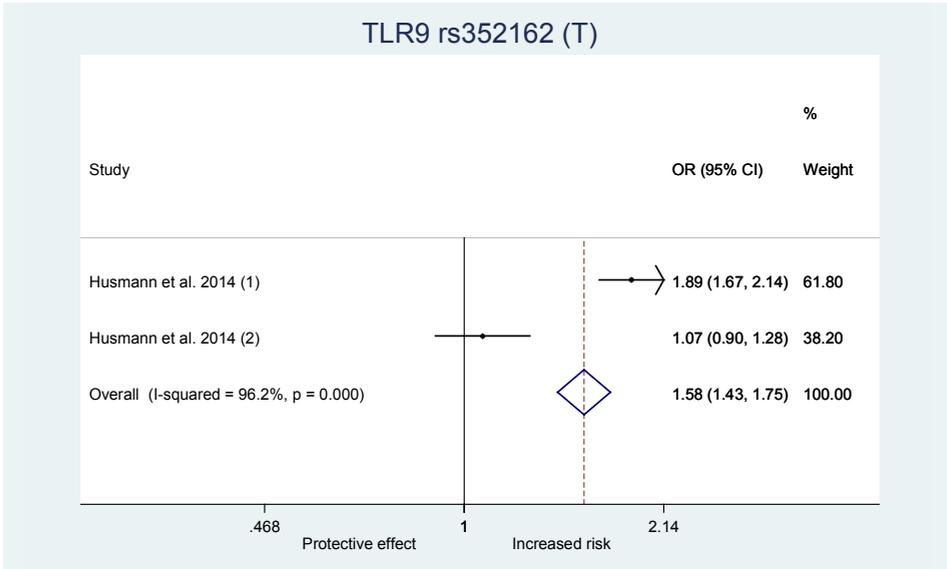
RXR_B rs6531 (C) forest plot. Harbold test: $p=0.292$, Egger test: $p=0.301$.
References: ⁵², ³, ²⁵



RXRB rs9277935 (T) forest plot. Harbold test: $p=0.393$, Egger test: $p=0.406$.
References: ⁵³, ³, ²⁵

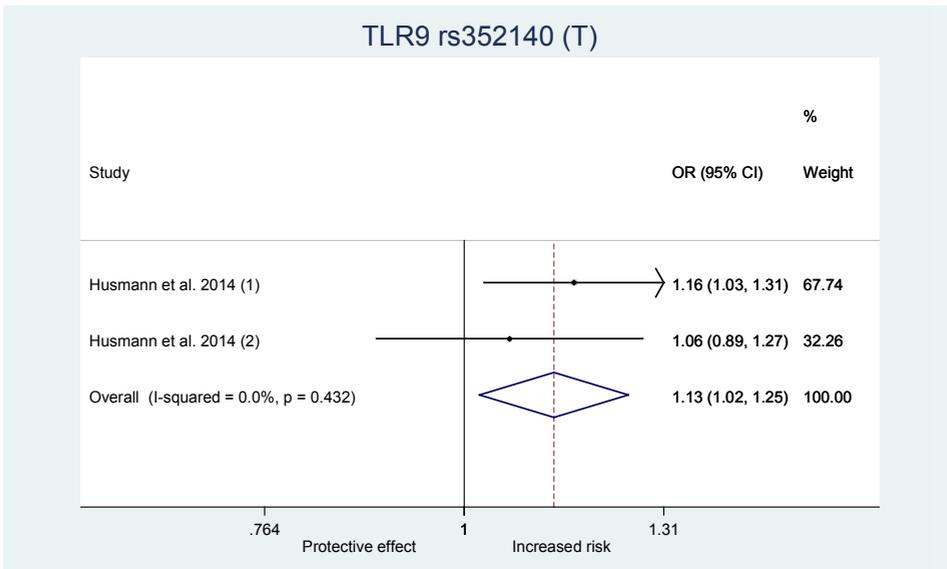


STAT4 rs7574865 (T) forest plot. Harbold test: $p=0.590$, Egger test: $p=0.567$.
References: ⁴⁴, ⁴⁵, ³



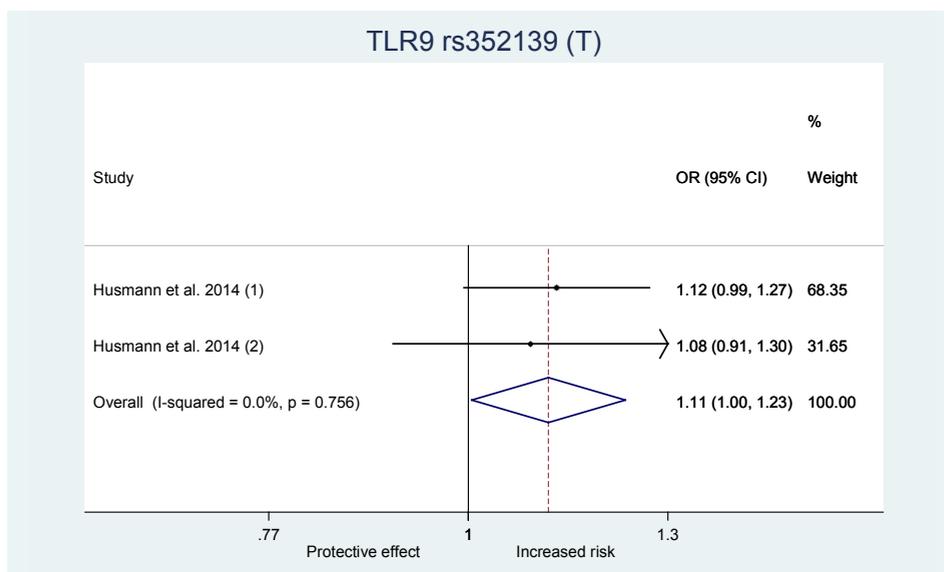
TLR9 rs352162 (T) forest plot. Harbold test: N/A, Egger test: N/A.

References: ^{40**}



TLR9 rs352140 (T) forest plot. Harbold test: N/A, Egger test: N/A.

References: ^{40**}



TLR9 rs352139 (T) forest plot. Harbold test: N/A, Egger test: N/A.

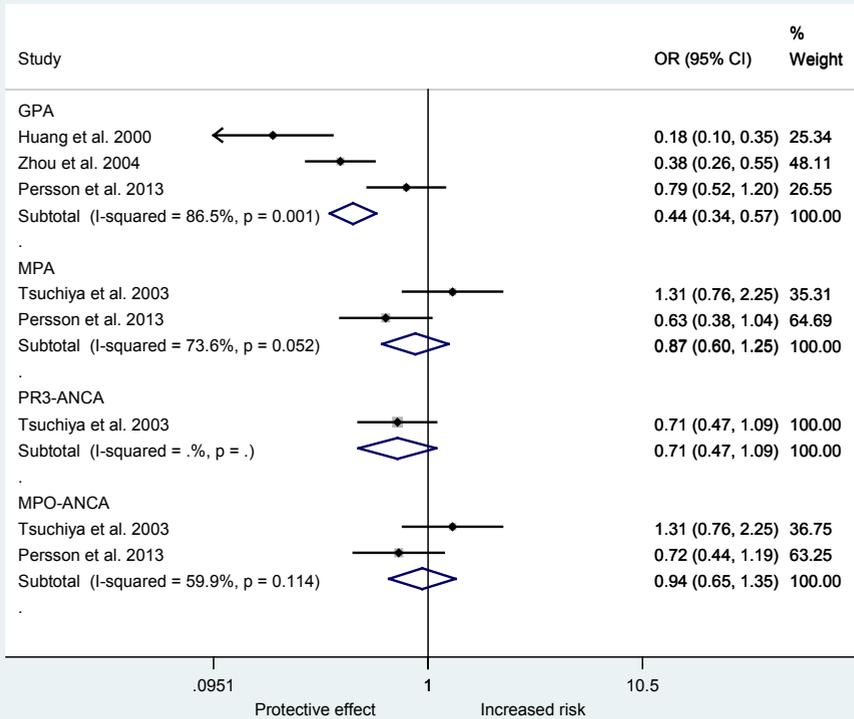
References: ⁴⁰**

**Two cohorts described in the same publication.

***Three cohorts described in the same publication.

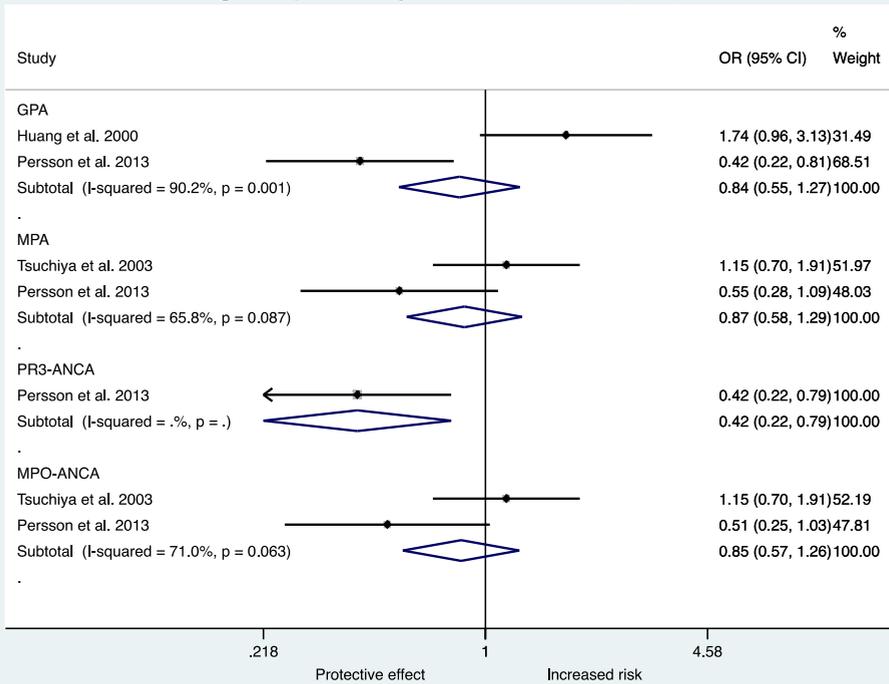
N/A, not applicable

Subgroup analysis CTLA-4 (AT)86



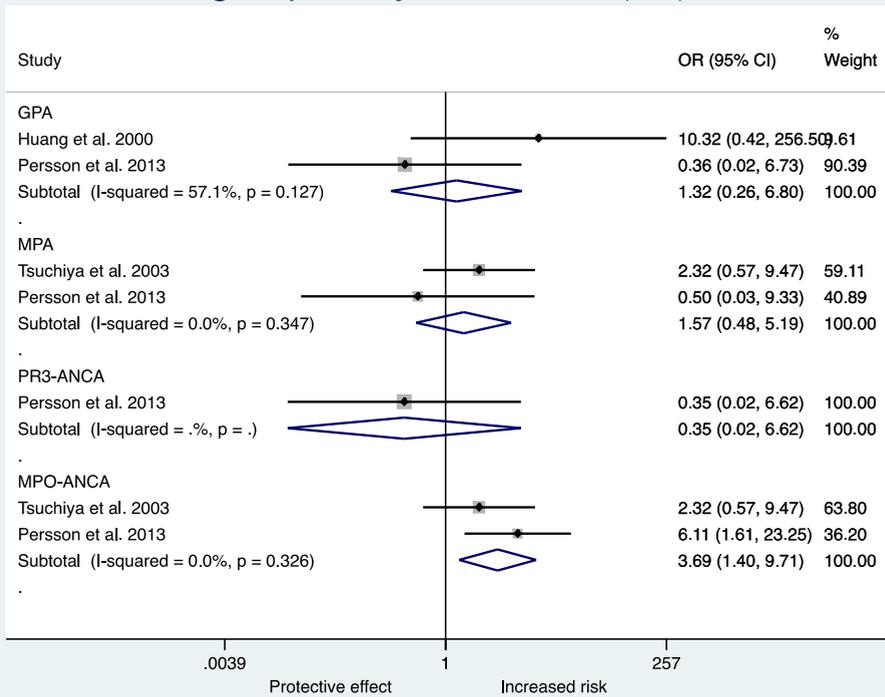
References: ⁴, ⁵, ⁶, ⁷

Subgroup analysis CTLA-4 (AT)104



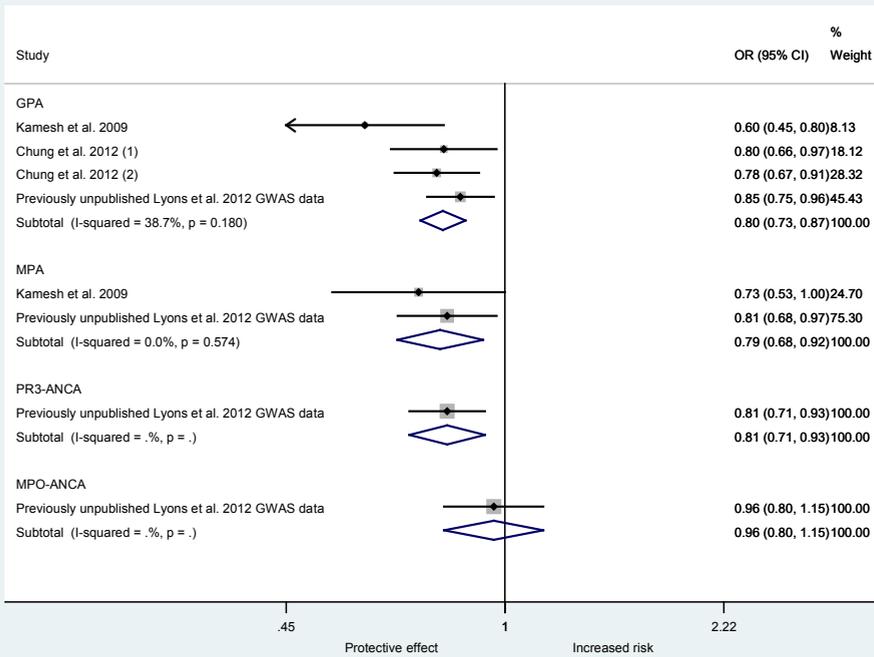
References: ⁴, ⁷, ⁵

Subgroup analysis CTLA-4 (AT)122



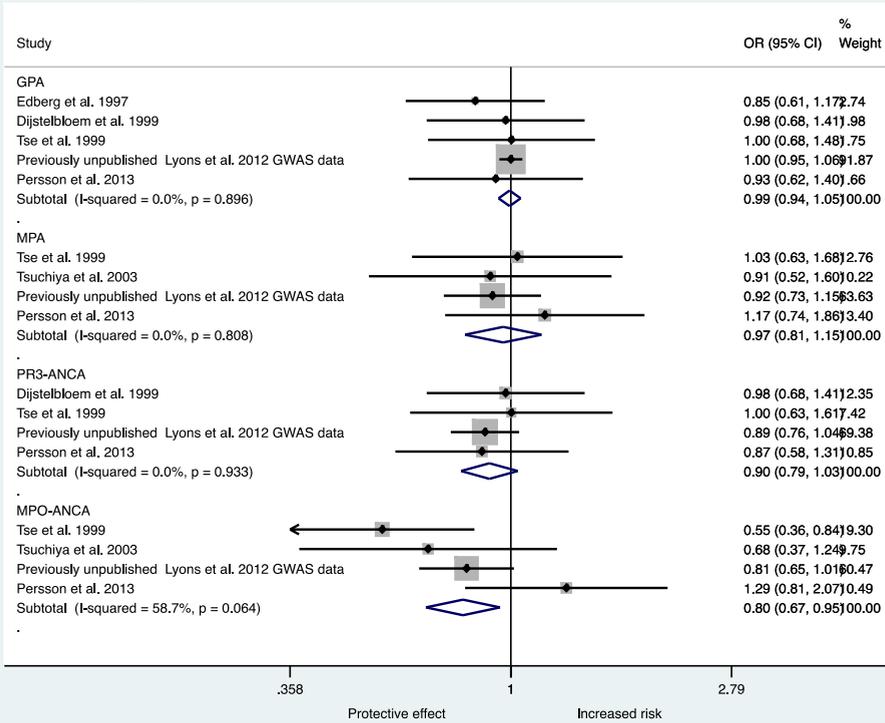
References: ⁴, ⁷, ⁵

Subgroup analysis CTLA-4 rs3087243 (A)



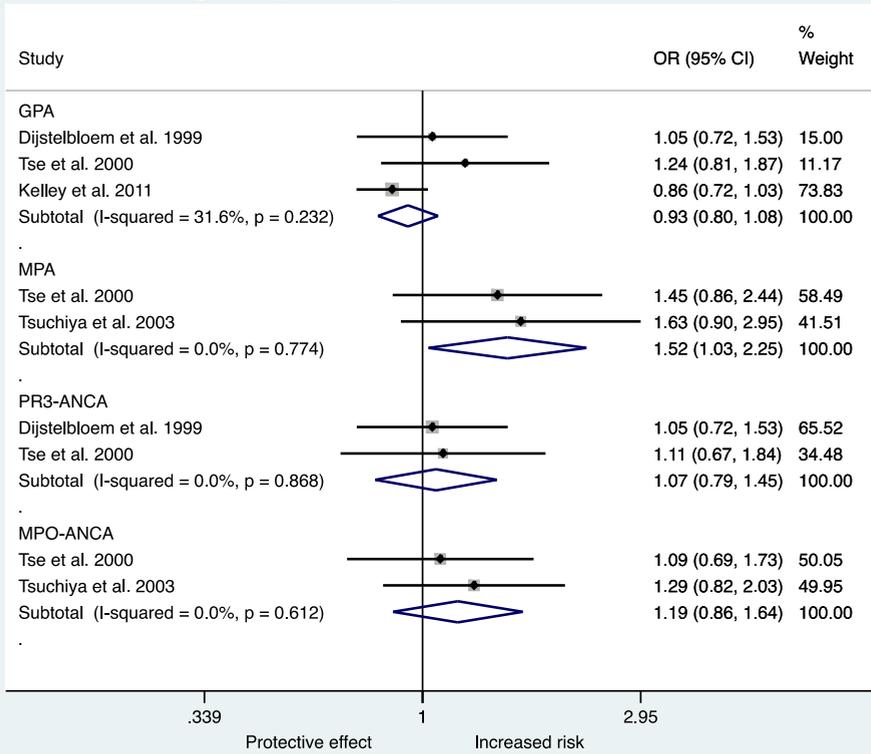
References: ⁹, ^{2**}, ³

Subgroup analysis FCGR2A rs1801274 (C)



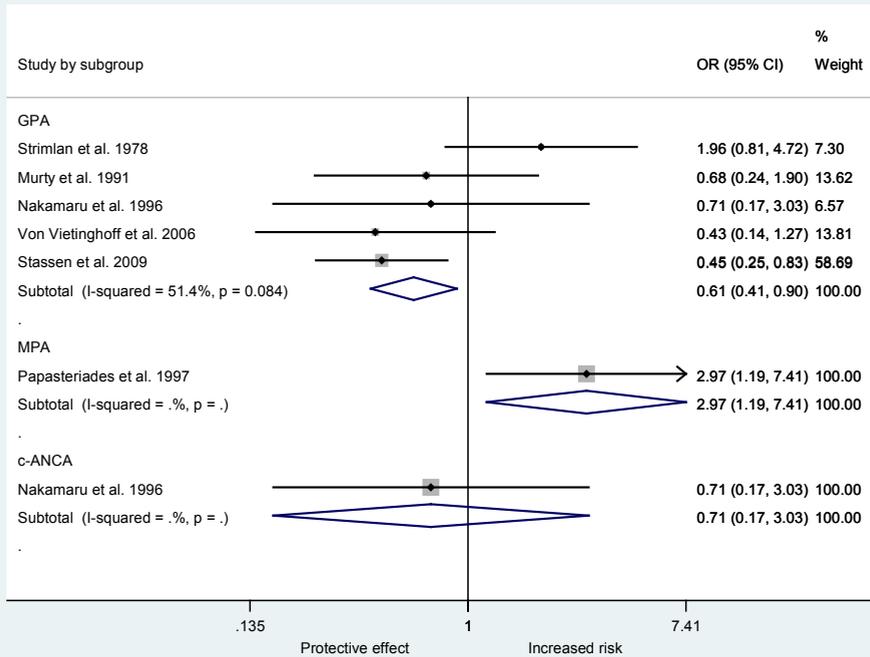
References: ¹¹, ¹², ¹³, ⁵, ⁷, ³

Subgroup analysis FCGR3B (NA1)



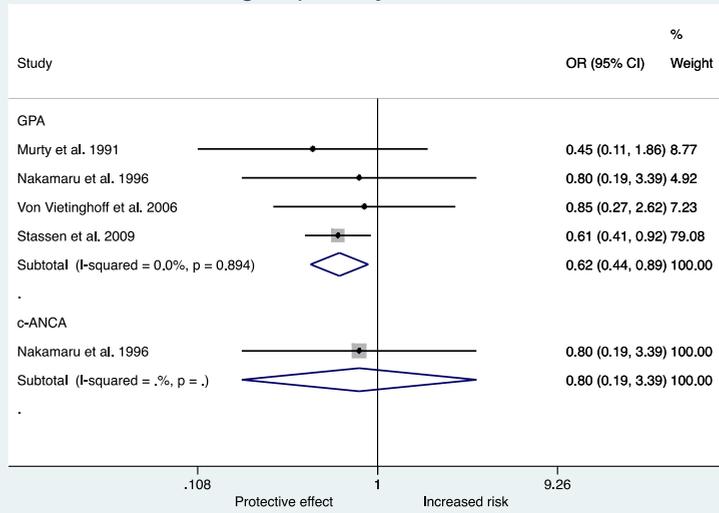
References: ¹², ¹⁴, ¹⁰, ⁵

Subgroup analysis HLA-A11



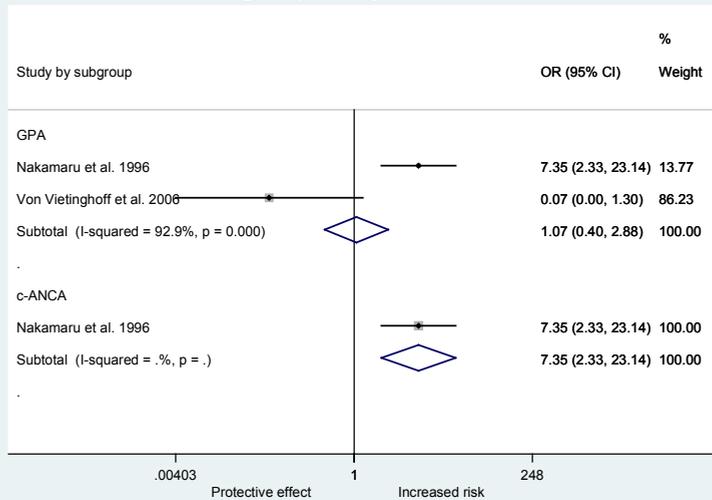
References: ¹⁶, ¹⁷, ²⁰, ²², ¹⁸, ¹⁹

Subgroup analysis HLA-B35



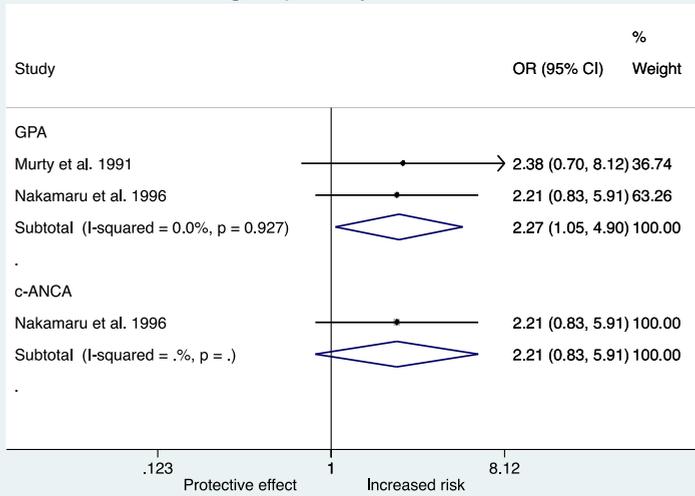
References: ¹⁷, ²⁰, ¹⁸, ¹⁹

Subgroup analysis HLA-B55



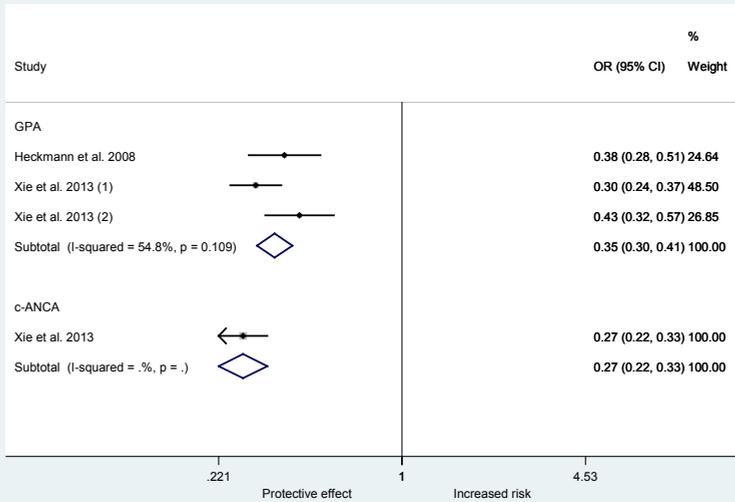
References: ²⁰, ¹⁸

Subgroup analysis HLA-B62



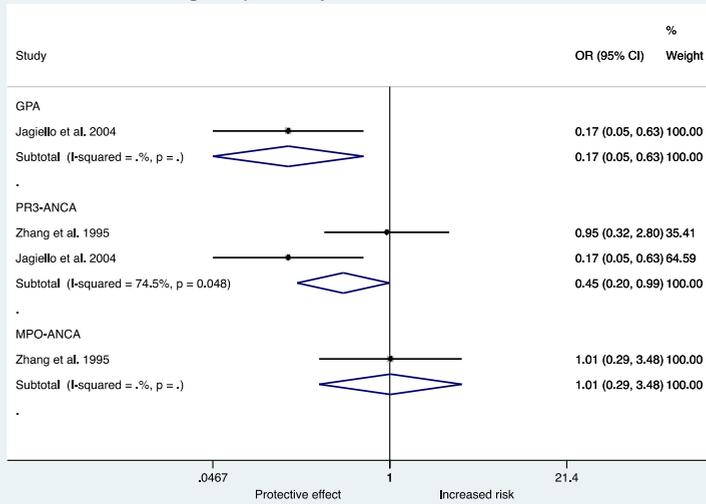
References: ^{17, 20}

Subgroup analysis HLA-DPA1 rs9277341 (C)



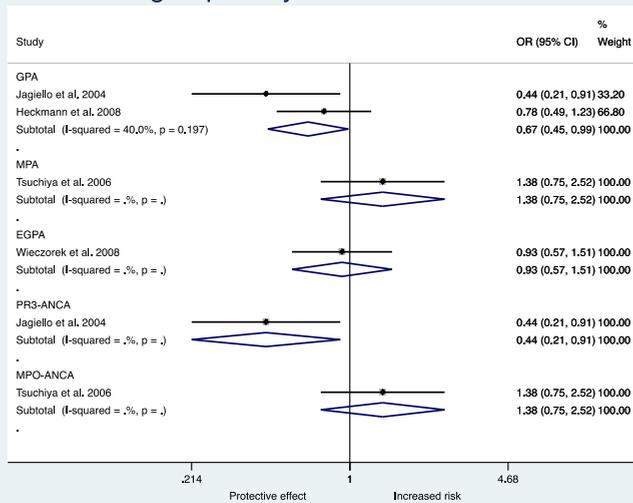
References: ^{24, 25**}

Subgroup analysis HLA-DPB1*0101



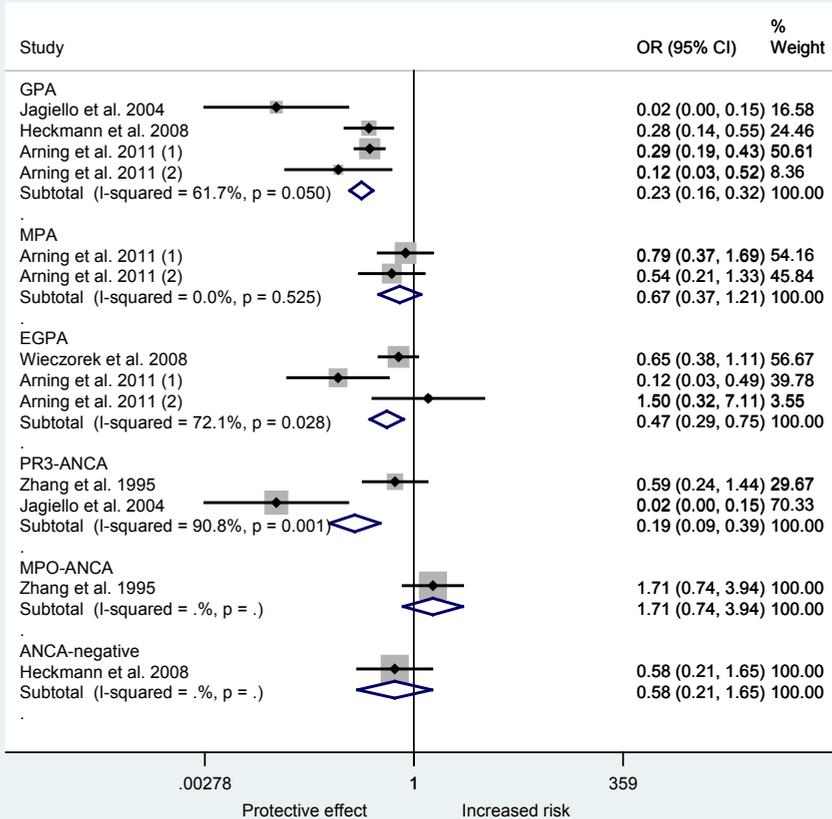
References: 27, 26

Subgroup analysis HLA-DPB1*0201



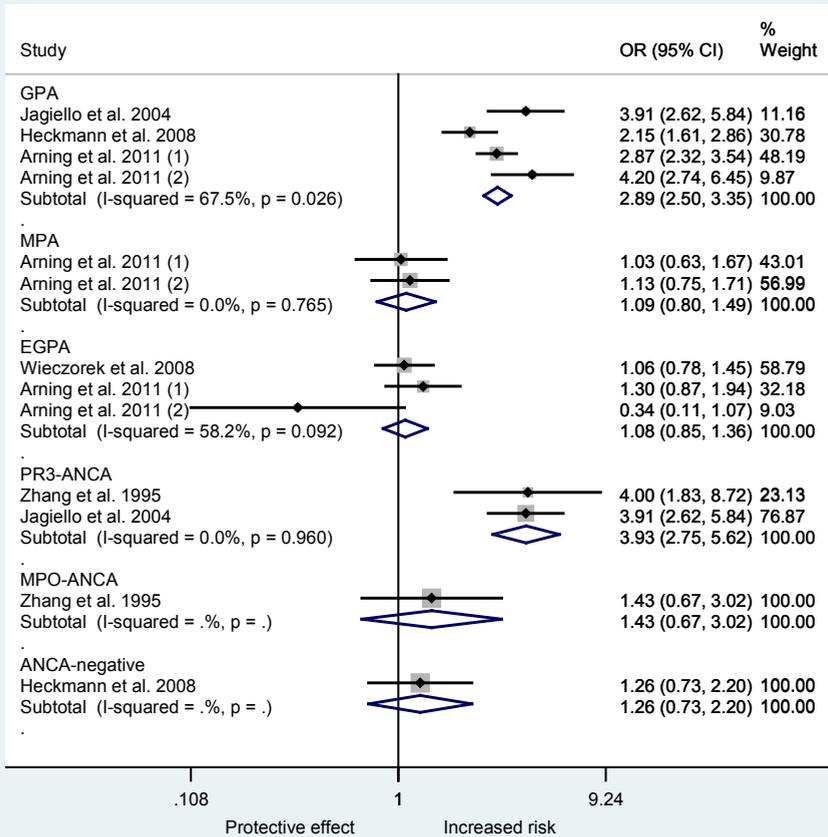
References: 27, 24, 28, 29

Subgroup analysis HLA-DPB1*0301



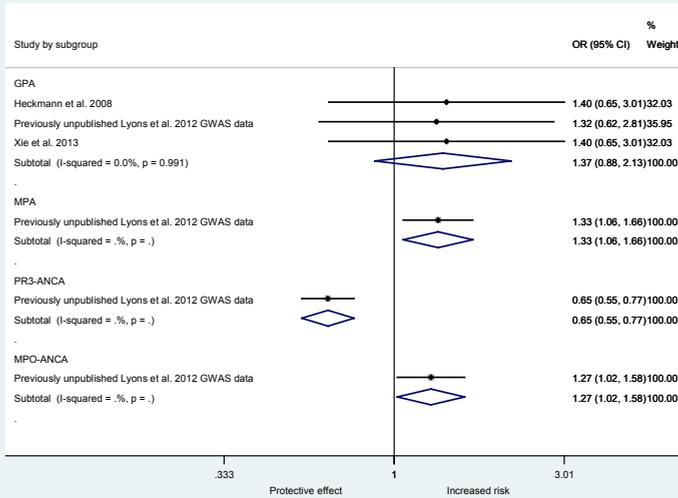
References: ²⁶, ²⁷, ²⁴, ²⁹, ^{30**}

Subgroup analysis HLA-DPB1*0401



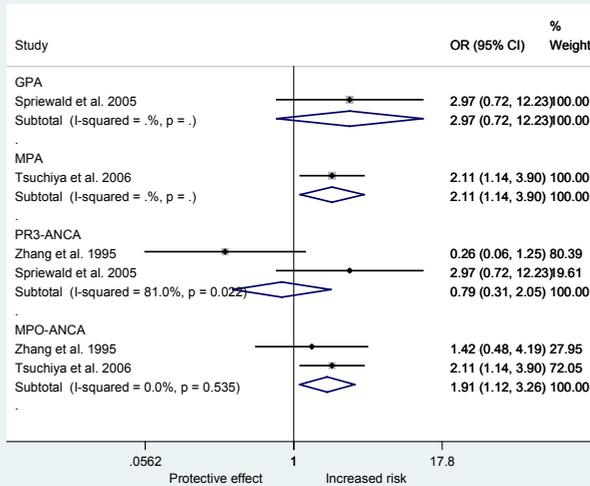
References: ²⁶, ²⁷, ²⁴, ²⁹, ^{30**}

Subgroup analysis HLA-DPB2 rs3130215 (A)



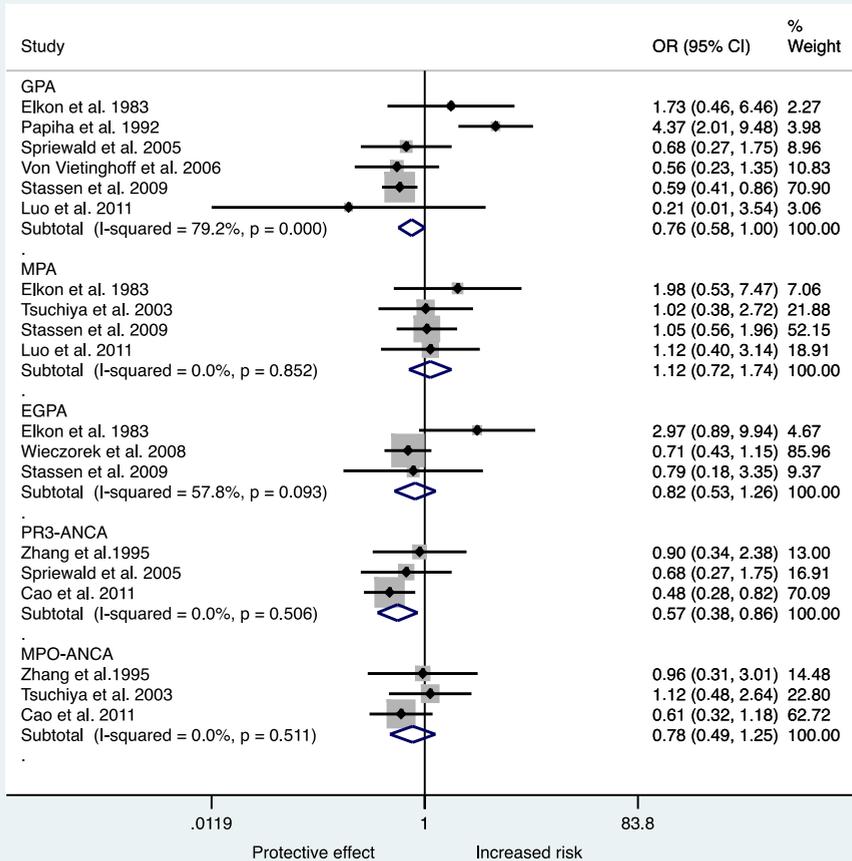
References: ²⁴, ³, ²⁵

Subgroup analysis HLA-DQB1*0303



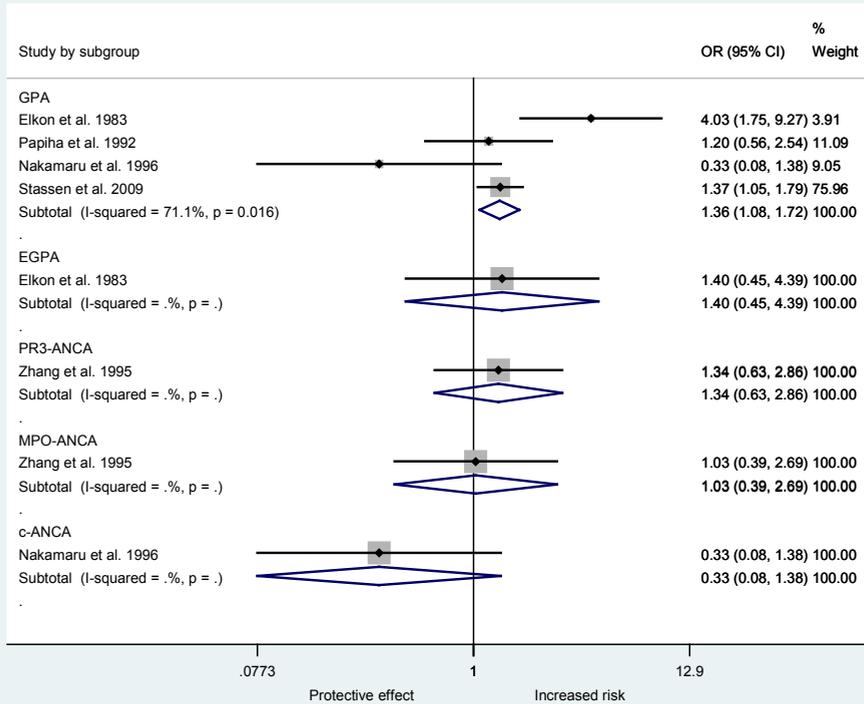
References: ²⁶, ³¹, ²⁸

Subgroup analysis HLA-DR1



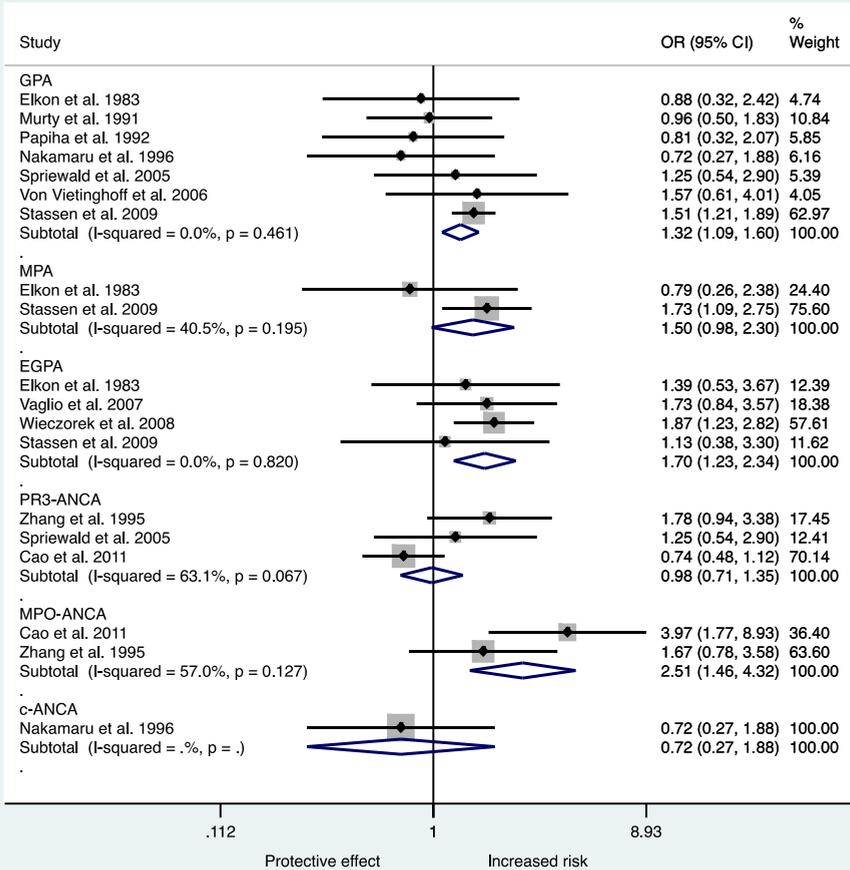
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Subgroup analysis HLA-DR2



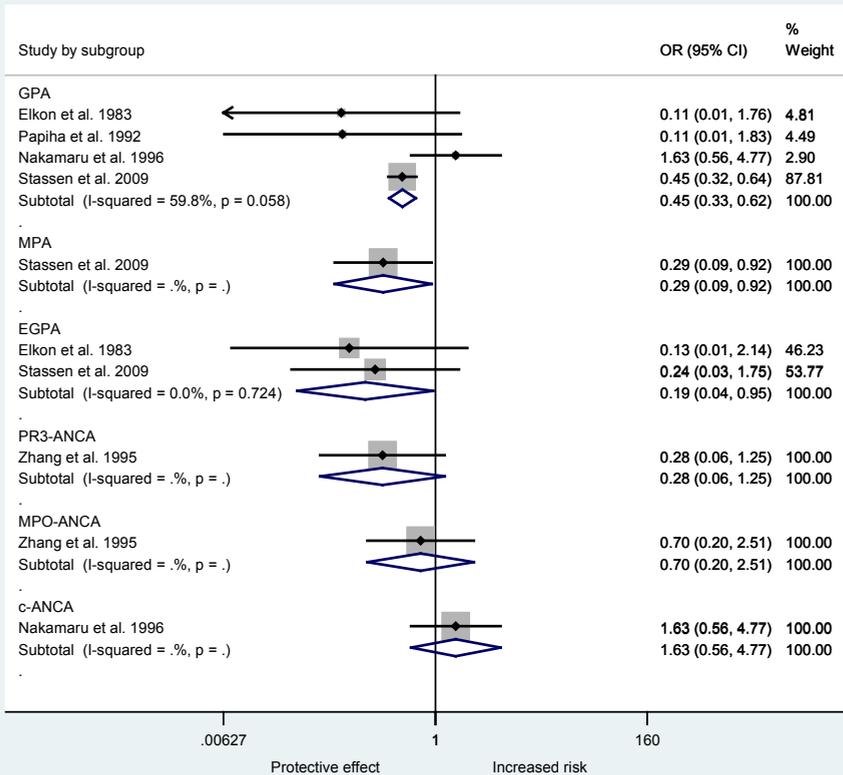
References: ²³, ³², ²⁶, ²⁰, ¹⁹

Subgroup analysis HLA-DR4



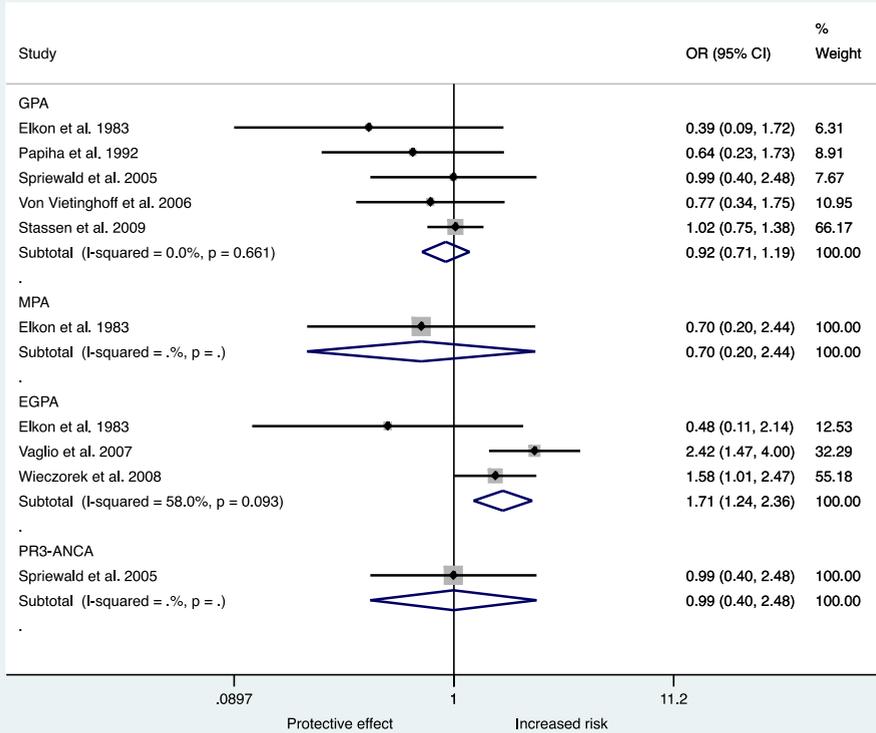
References: ²³, ¹⁷, ³², ²⁰, ³¹, ¹⁸, ¹⁹, ³⁵, ²⁹, ²⁶, ³³

Subgroup analysis HLA-DR6



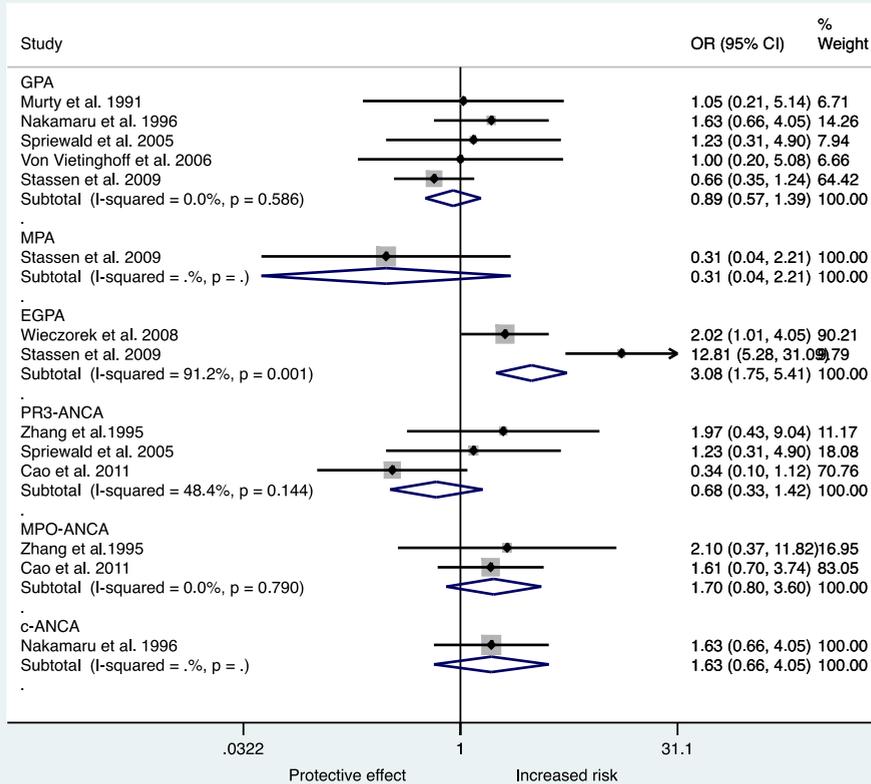
References: ²³, ³², ²⁶, ²⁰, ¹⁹

Subgroup analysis HLA-DR7



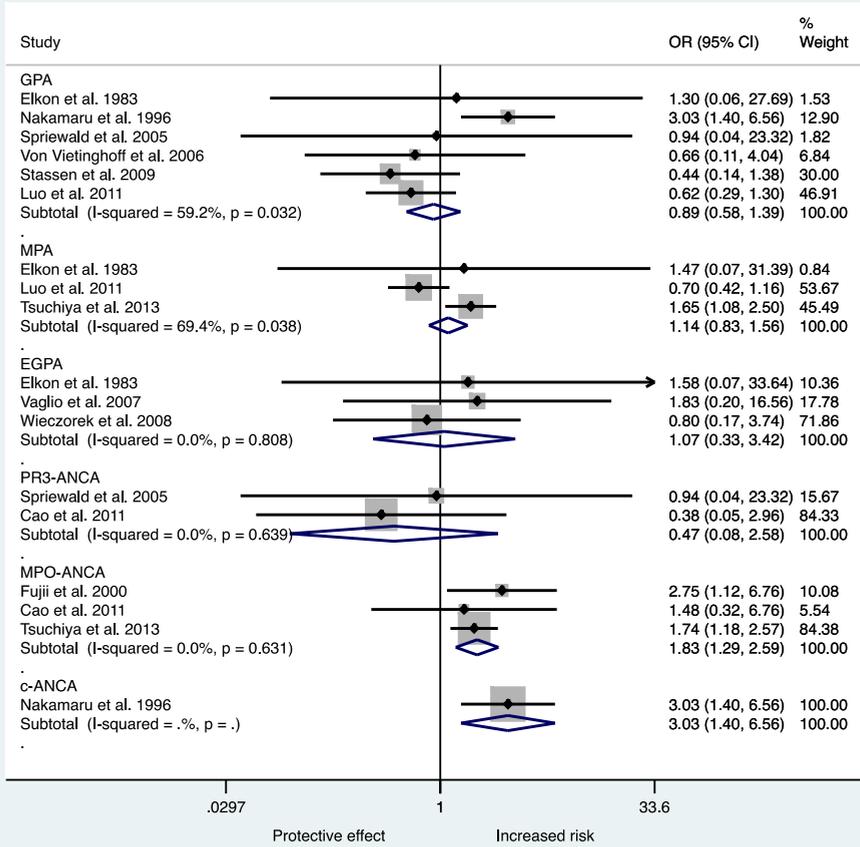
References: ²³, ³², ³¹, ¹⁸, ¹⁹, ³⁵, ²⁹

Subgroup analysis HLA-DR8



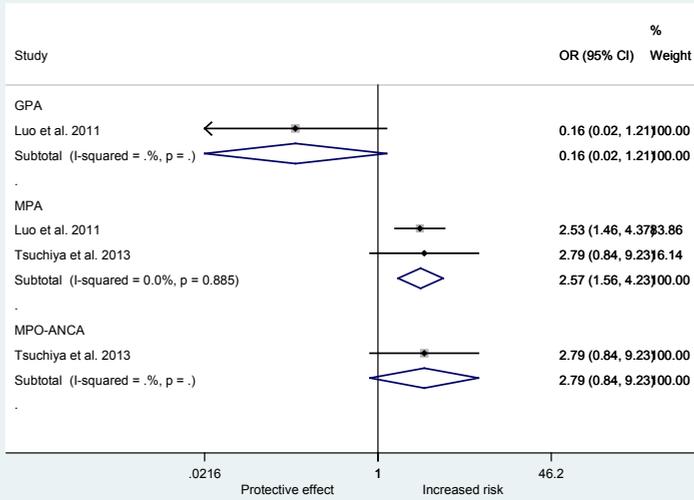
References: ¹⁷, ²⁰, ³¹, ¹⁸, ¹⁹, ²⁹, ²⁶, ³³

Subgroup analysis HLA-DR9



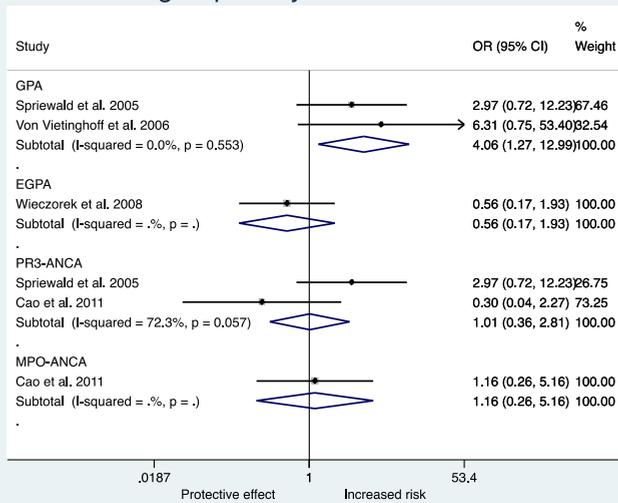
References: 23, 20, 31, 18, 19, 34, 37, 35, 29, 33, 36

Subgroup analysis HLA-DRB1*1101



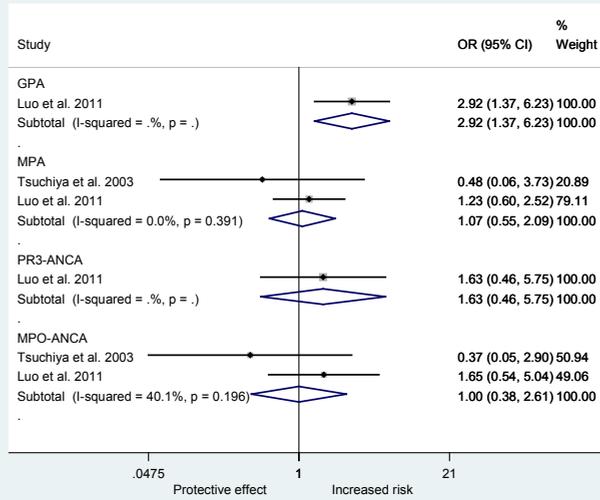
References: ³⁴, ³⁷

Subgroup analysis HLA-DRB1*12



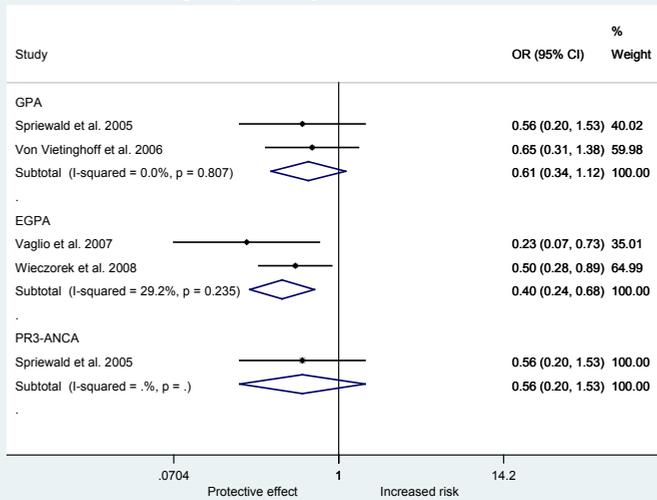
References: ³¹, ¹⁸, ²⁹, ³³

Subgroup analysis HLA-DRB1*1202



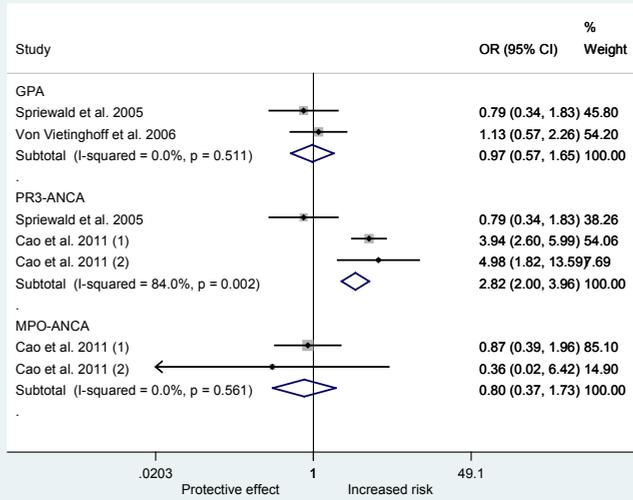
References: ^{5, 34}

Subgroup analysis HLA-DRB1*13



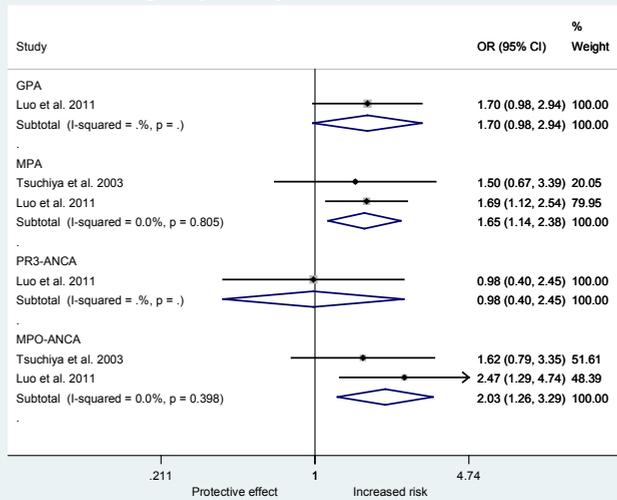
References: ^{31, 18, 35, 29}

Subgroup analysis HLA-DRB1*15



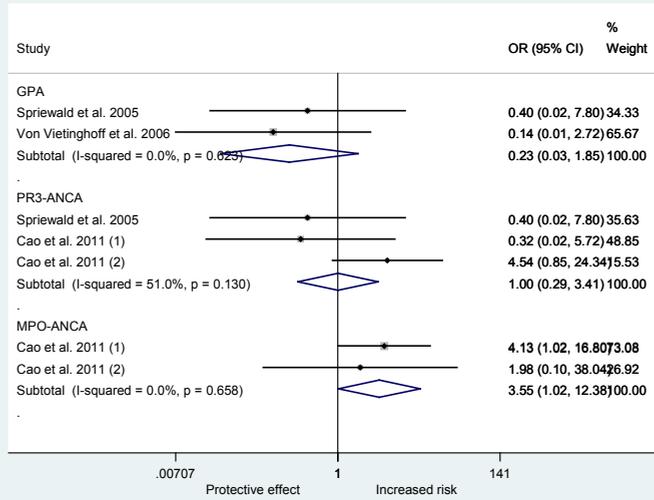
References: ^{31, 18, 33**}

Subgroup analysis HLA-DRB1*1501



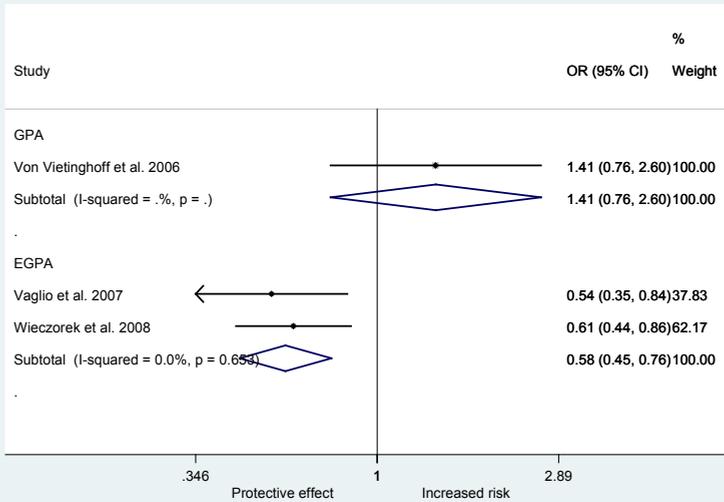
References: ^{5, 34}

Subgroup analysis HLA-DRB1*16



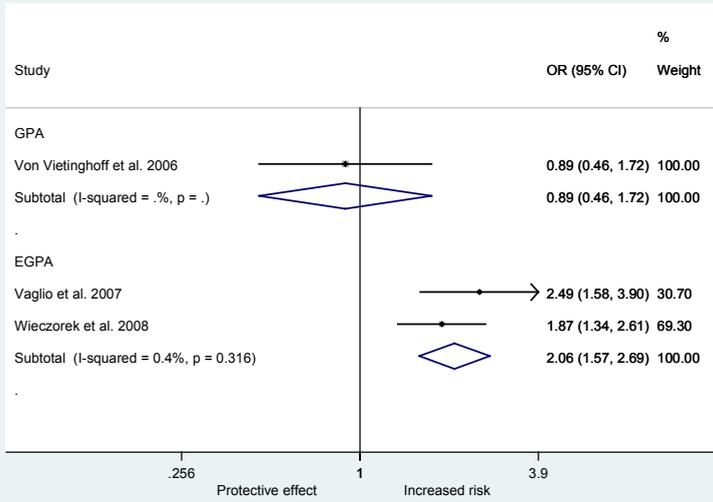
References: ³¹, ¹⁸, ³³**

Subgroup analysis HLA-DRB3



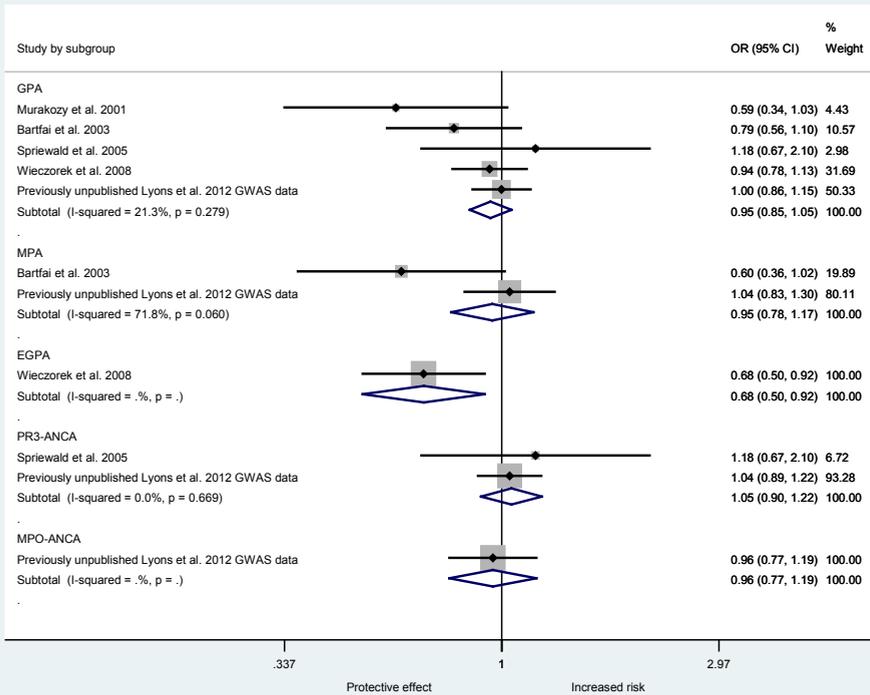
References: ¹⁸, ³⁵, ²⁹

Subgroup analysis HLA-DRB4



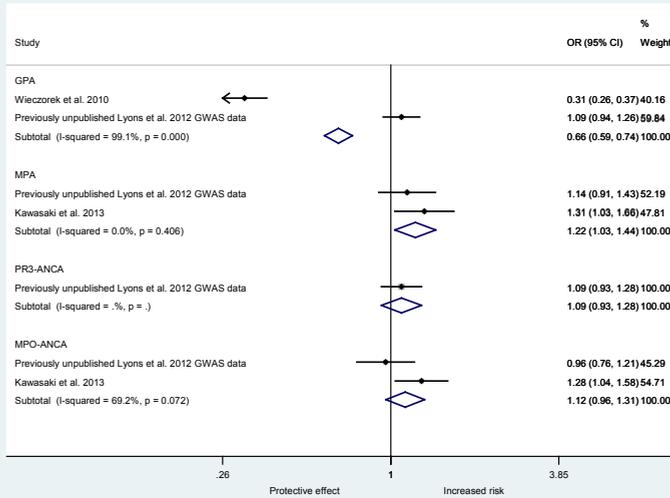
References: ^{18, 35, 29}

Subgroup analysis IL-10 rs1800896 (G)



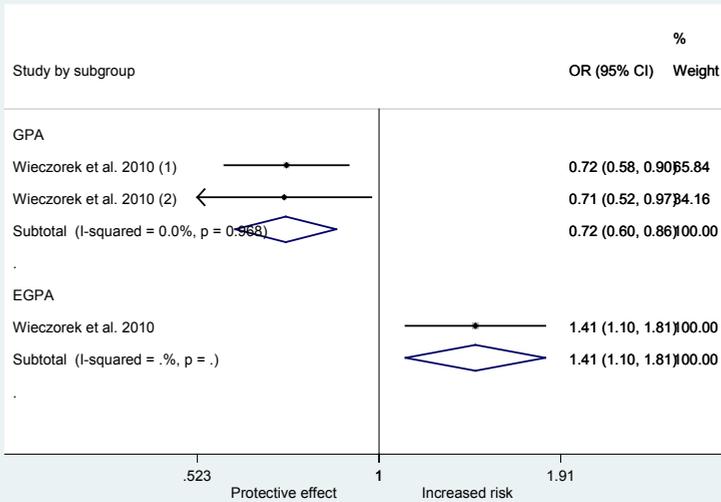
References: ⁴², ⁴³, ³¹, ⁴¹, ³

Subgroup analysis IRF5 rs10954213 (G)



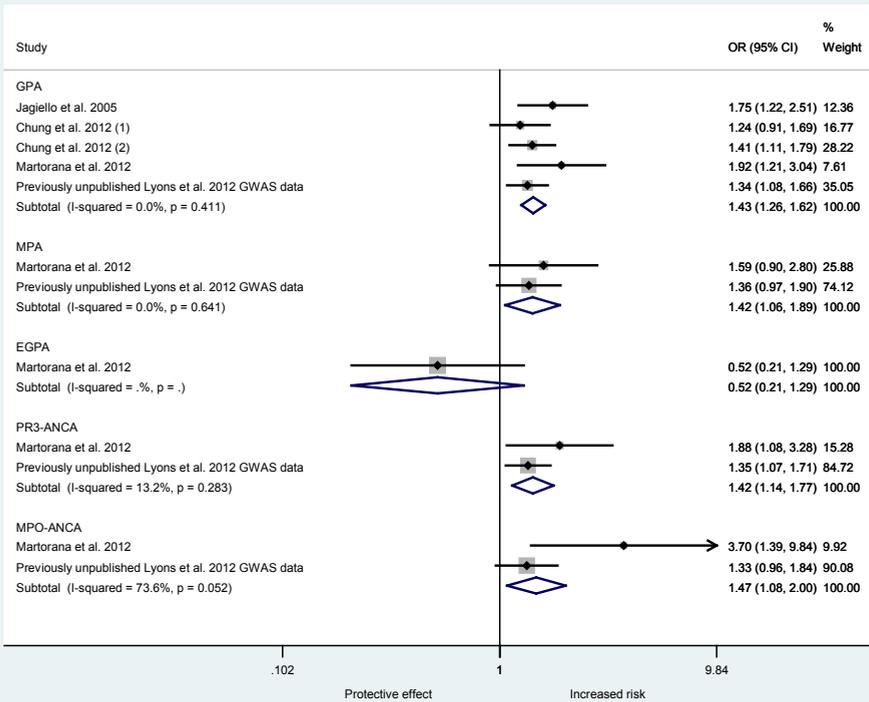
References: ⁴⁴, ³, ⁴⁵

Subgroup analysis LEPR rs8179183 (C)



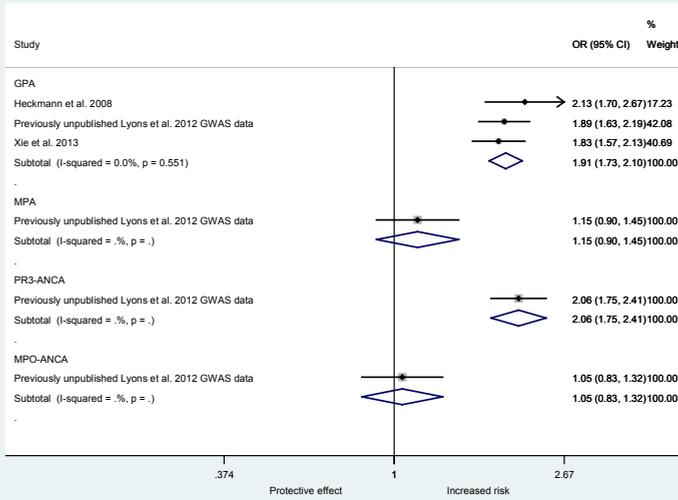
References: ¹⁵**

Subgroup analysis PTPN22 rs2476601 (A)



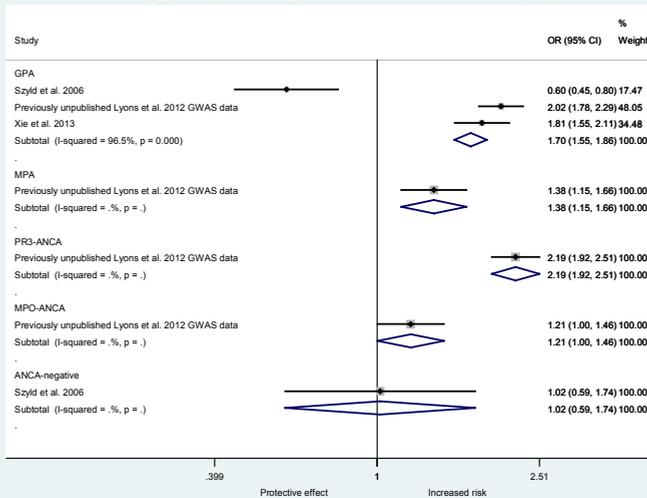
References: ⁵⁰, ^{2**}, ⁵¹, ³

Subgroup analysis RING1/RXRB rs213213 (A)



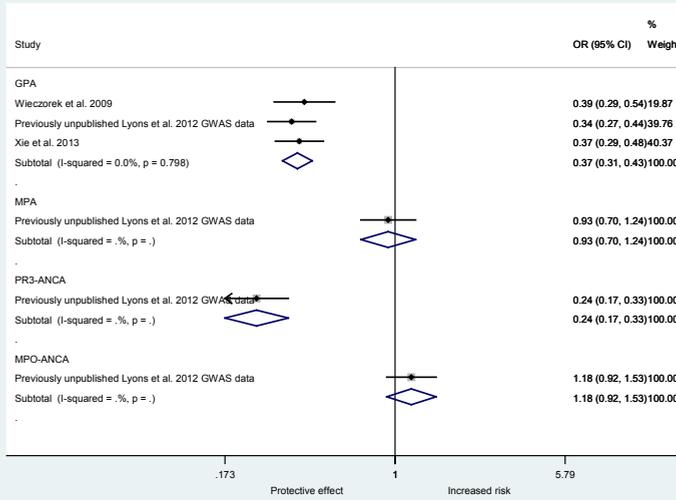
References: ²⁴, ³, ²⁵

Subgroup analysis RXRB rs6531 (C)



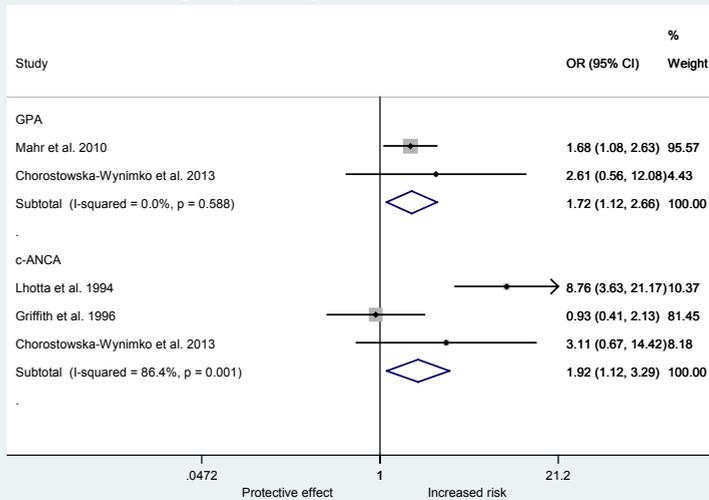
References: ⁵², ³, ²⁵

Subgroup analysis RXRB rs9277935 (T)



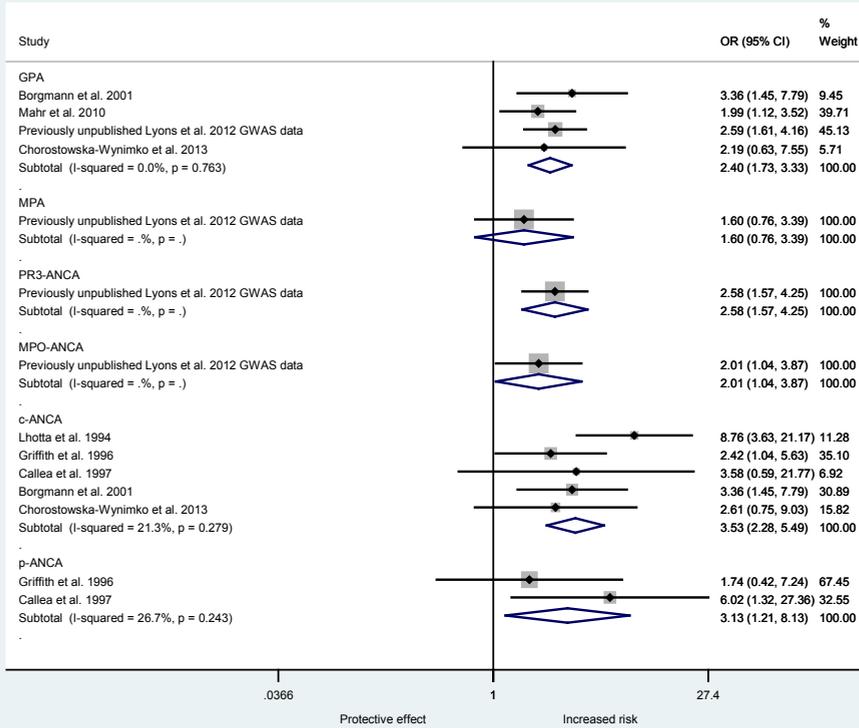
References: ⁵³, ³, ²⁵

Subgroup analysis SERPINA1 S allele



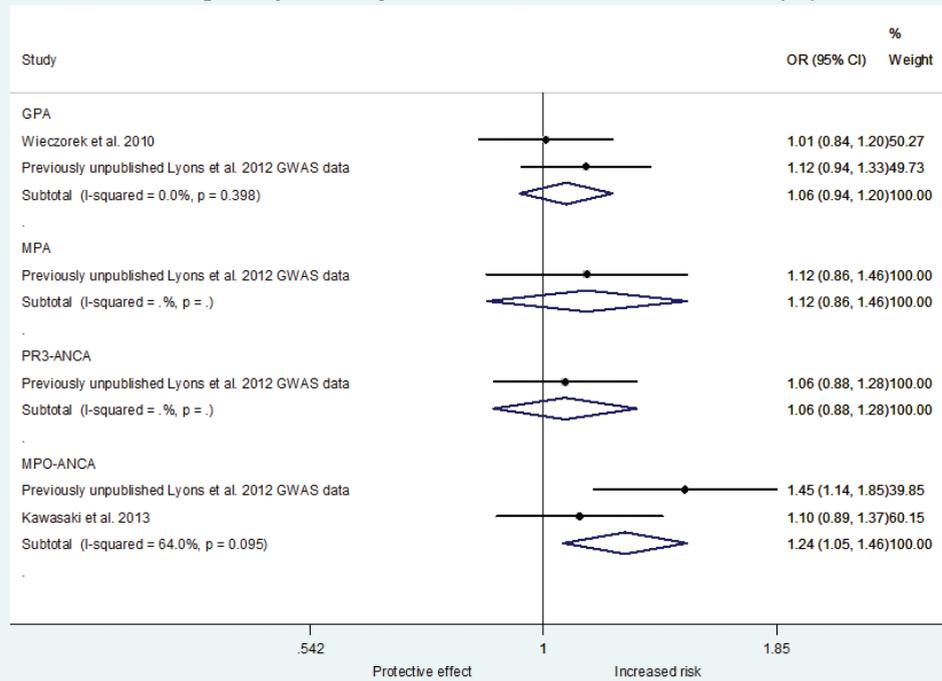
References: ⁵⁴, ⁵⁵, ⁵⁶, ⁵⁸

Subgroup analysis SERPINA1 Z allele



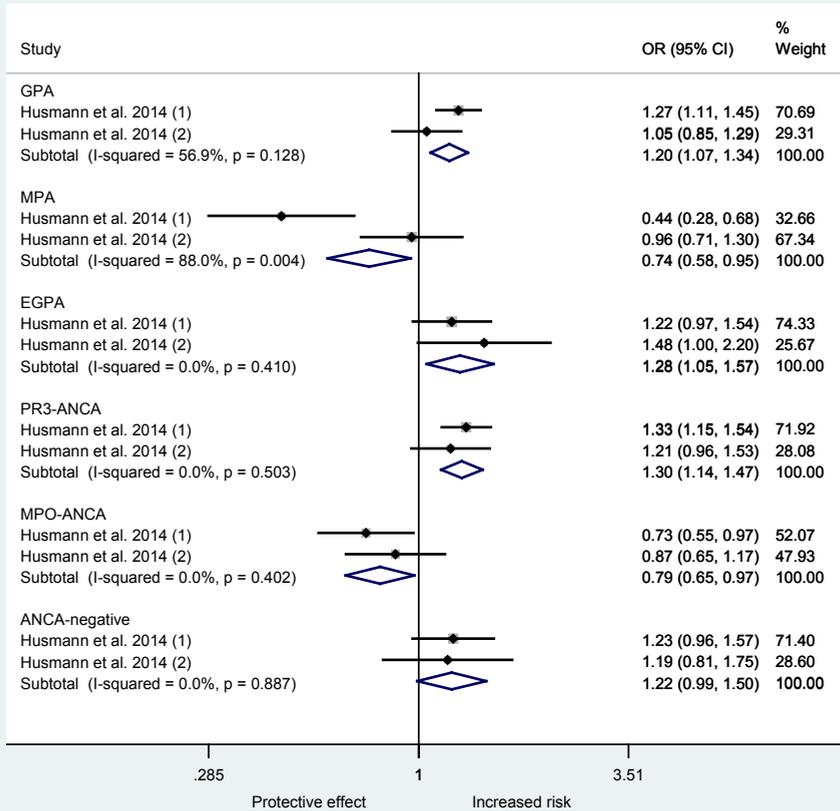
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Subgroup analysis STAT4 rs7574865 (T)



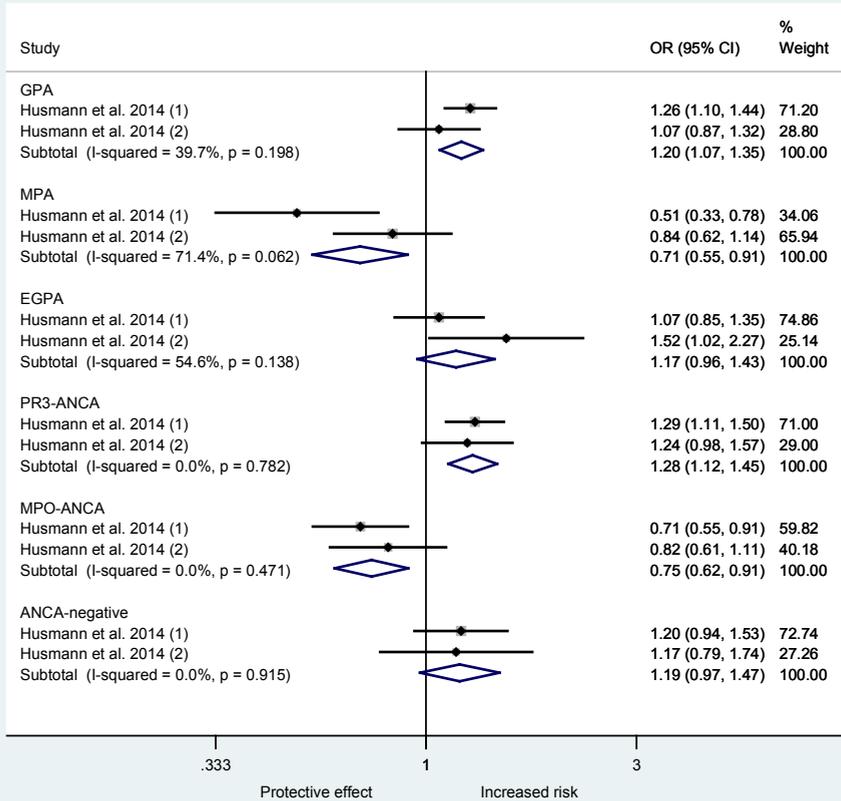
References: ⁴⁴, ⁴⁵

Subgroup analysis TLR9 rs352162 (T)



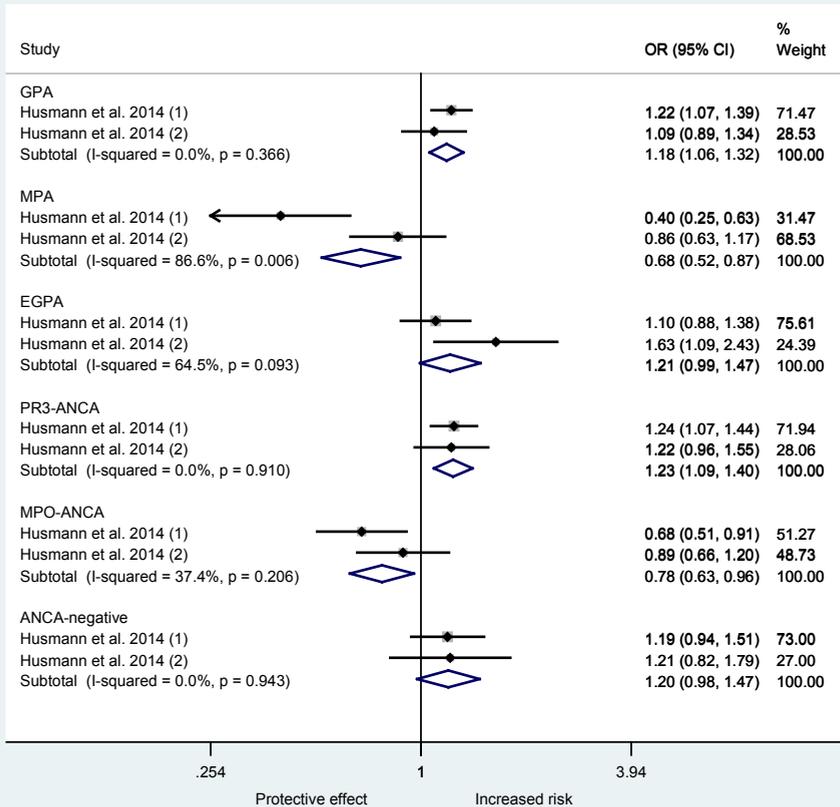
References: ^{40**}

Subgroup analysis TLR9 rs352140 (T)



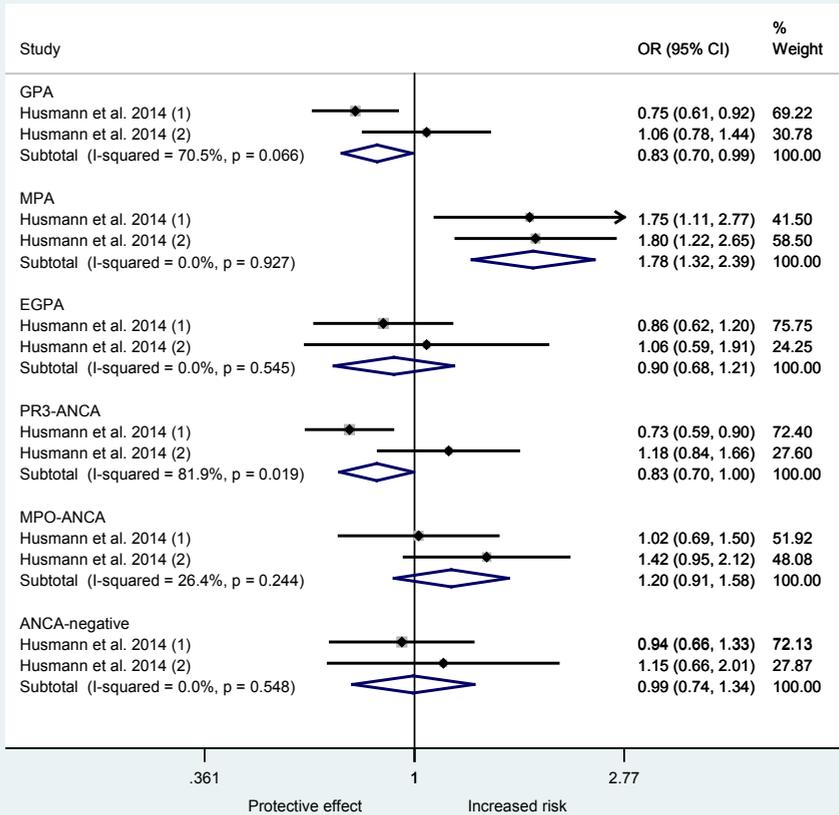
References: ⁴⁰**

Subgroup analysis TLR9 rs352139 (T)



References: ^{40**}

Subgroup analysis TLR9 rs5743836 (G)

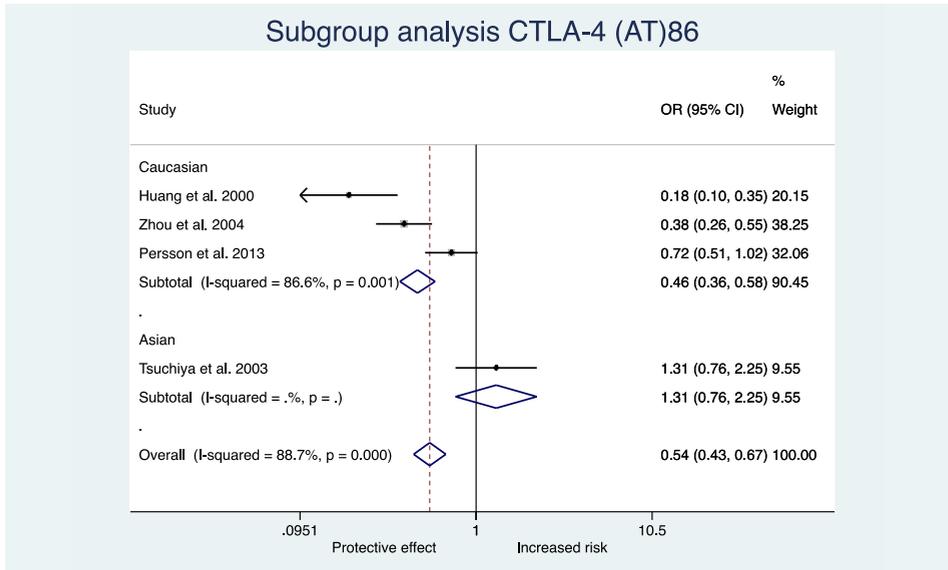


References: ⁴⁰**

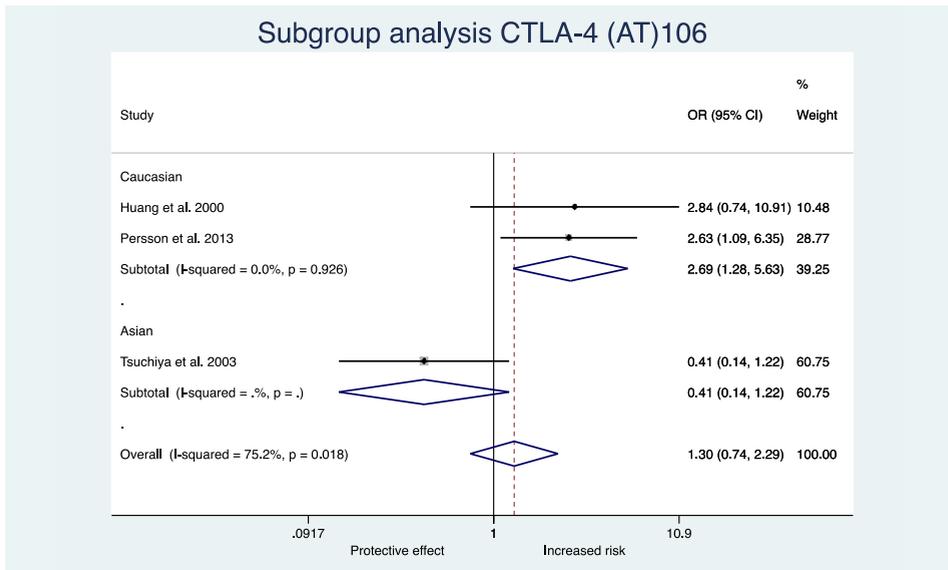
**Two cohorts described in the same publication.

***Three cohorts described in the same publication.

Supplementary figure 3. Forest plots by ethnic subgroups

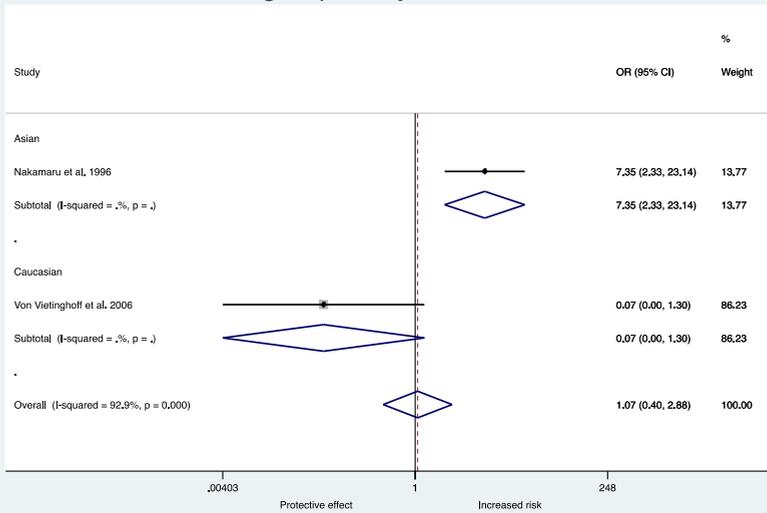


References: ⁴, ⁵, ⁶, ⁷



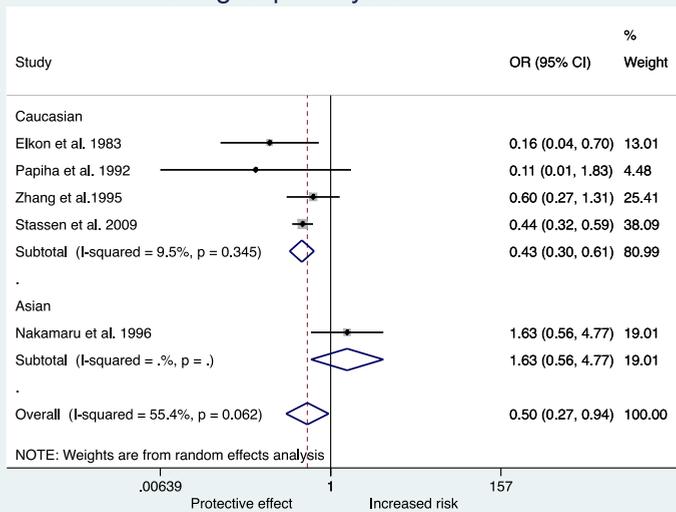
References: ⁴, ⁵, ⁷

Subgroup analysis HLA-B55



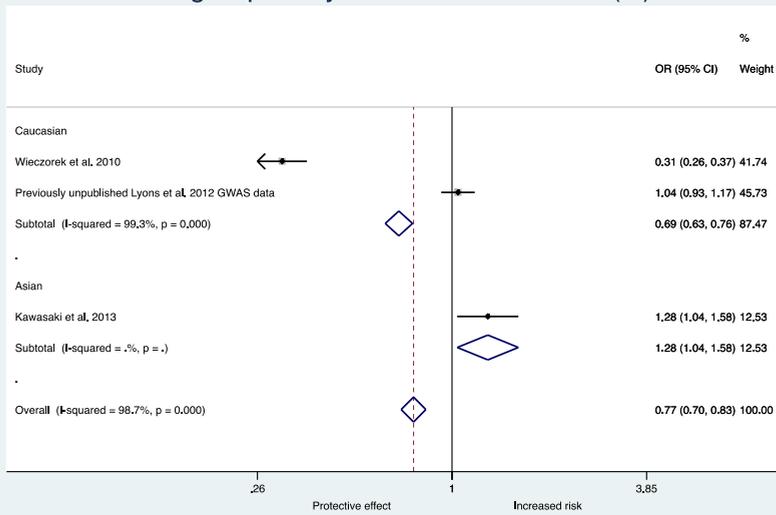
References: ^{20, 18}

Subgroup analysis HLA-DR6



References: ^{23, 32, 26, 20, 19}

Subgroup analysis IRF5 rs10954213 (G)



References: ⁴⁴, ³, ⁴⁵

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II. Supplementary data Chapter III

Renal function and ear, nose, throat involvement in ANCA associated vasculitis: prospective data from the European Vasculitis Society clinical trials

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Supplementary table 1. ENT manifestations

ENT symptom	Patients
Nasal obstruction	76 (43)
Bloody nasal discharge	70 (40)
Nasal crusting	60 (34)
Sinus involvement	45 (25)
Hearing loss	46 (26)
Hoarseness/stridor	12 (7)
Otorhinolaryngologist's opinion	
Granulomatous sinusitis	28 (16)
Conductive hearing loss	20 (11)
Sensorineural hearing loss	9 (5)
Significant subglottic inflammation	4 (2)

All data are presented as n (%). ENT symptoms scored using the Birmingham Vasculitis Activity Score are shown. Data were available for 177 of the 185 patients with ENT involvement. Percentages are expressed relative to the number of patients with ENT involvement. All items below 'otorhinolaryngologist's opinion' were only scored by the otorhinolaryngologist. ENT, ear-, nose-, and throat.

Supplementary table 2. Associations between ENT symptoms and ANCA-subtype in patients with ENT involvement

ENT symptom	PR3-ANCA patients (n=116)	MPO-ANCA patients (n=46)	P Value
Nasal obstruction	57 (49)	14 (30)	0.03
Bloody nasal discharge	45 (39)	19 (41)	0.77
Nasal crusting	43 (37)	13 (28)	0.29
Sinus involvement	30 (26)	13 (28)	0.76
Hearing loss	34 (29)	7 (15)	0.06
Hoarseness/stridor	7 (6)	5 (11)	0.30
Otorhinolaryngologist's opinion			
Granulomatous sinusitis	25 (22)	2 (4)	0.008
Conductive hearing loss	16 (14)	3 (7)	0.20
Sensorineural hearing loss	5 (4)	2 (4)	0.99
Significant subglottic inflammation	2 (2)	2 (4)	0.33

Data are presented as n (%). The distribution of ANCA-subtype in patients with ENT involvement is shown. Data regarding the specific ENT symptoms were available for 177 of the 185 patients with ENT involvement. All 8 patients with missing ENT symptoms data were PR3-ANCA positive. All items listed under 'otorhinolaryngologist's opinion' were scored by the otorhinolaryngologist alone. ENT, ear-, nose-, and throat; PR3-ANCA, anti-proteinase 3 anti-neutrophil cytoplasm antibody; MPO-ANCA, anti-myeloperoxidase anti-neutrophil cytoplasm antibody.

Supplementary table 3. Associations between ENT symptoms and baseline eGFR

ENT symptom	eGFR in symp- tom+ patients	eGFR in symp- tom- patients	95% CI	P Value
	(mL/min/1.73 m ²)	(mL/min/1.73 m ²)		
Nasal obstruction	43.09	26.59	9.91 – 23.10	<0.001
Bloody nasal discharge	37.81	28.05	2.82 – 16.71	0.006
Nasal crusting	43.25	27.36	8.60 – 23.19	<0.001
Sinus involvement	36.38	28.93	-0.95 – 15.84	0.08
Hearing loss	36.78	28.86	-0.40 – 16.23	0.06
Hoarseness/stridor	28.70	29.82	-16.72 – 14.47	0.89
Otorhinolaryngologist's opinion				
Granulomatous sinusitis	41.56	28.88	2.32 – 23.03	0.02
Conductive hearing loss	44.89	29.98	3.80 – 28.02	0.01
Sensorineural hearing loss	26.88	29.86	-20.91 – 14.96	0.74
Significant subglottic inflammation	45.53	29.63	-10.78 – 42.60	0.24

ENT symptoms were scored using the Birmingham Vasculitis Activity Score. Data regarding the specific ENT symptoms were available for 177 of the 185 patients with ENT involvement. All items listed under 'otorhinolaryngologist's opinion' were scored by the otorhinolaryngologist alone. ENT, ear-, nose-, and throat; eGFR, estimated glomerular filtration rate.

Supplementary table 4. Correlations of clinical and histological parameters with baseline and 5-year follow-up eGFR

	Baseline eGFR		5-year follow-up eGFR	
	r	p Value	r	P Value
ENT involvement	0,274	< 0.001	0,224	0.005
Age	-0,379	< 0.001	-0,395	< 0.001
PR3-ANCA	0,240	< 0.001	0,097	<0.24
Tubulitis	-0,445	< 0.001	-0,266	0.12
Interstitial infiltrate	-0,477	< 0.001	-0,396	< 0.001
IFTA	-0,436	< 0.001	-0,477	< 0.001
AAGN classification	-0,424	< 0.001	-0,511	< 0.001

ENT, ear-, nose-, and throat; PR3-ANCA, anti-proteinase 3 anti-neutrophil cytoplasm antibody; IFTA, interstitial fibrosis and tubular atrophy; AAGN, anti-neutrophil cytoplasm antibody associated glomerulonephritis; eGFR, estimated glomerular filtration rate.

Supplementary table 5. Three multivariable regression models investigating the association between ENT involvement and baseline eGFR

Model 1	β (95% CI)	P Value
ENT involvement	10.40 (5.24 – 15.55)	< 0.001
Age	-0.65 (-0.82 – -0.47)	< 0.001
PR3-ANCA	6.21 (1.03 – 11.38)	0.02
Model 2	β (95% CI)	P Value
ENT involvement	10.32 (4.05 – 16.59)	0.001
Age	-0.46 (-0.68 – -0.24)	< 0.001
PR3-ANCA	-1.43 (-7.73 – 4.88)	0.66
Tubulitis	-13.93 (-20.87 – -6.99)	< 0.001
Interstitial infiltrate	-8.56 (-12.64 – -4.47)	< 0.001
IFTA	-10.54 (-15.61 – -5.47)	< 0.001
Model 3	β (95% CI)	P Value
ENT involvement	9.14 (2.30 – 15.98)	0.009
Age	-0.60 (-0.83 – -0.36)	< 0.001
PR3-ANCA	-1.26 (-8.28 – 5.75)	0.71
Tubulitis	-17.82 (-25.78 – -9.86)	< 0.001
Interstitial infiltrate	-6.86 (-11.20 – -2.52)	0.002
IFTA	-8.72 (-14.67 – -2.77)	0.004
AAGN classification	-6.20 (-9.84 – -2.57)	0.001

Since renal biopsies were not available for all patients, including the histopathological parameters limited the number of patients included in the analysis. To be able to include all patients we therefore created three models. The first model included only clinical parameters (n=412). In the second model, we added tubulointerstitial parameters (n=195). The third model, we added the histopathological classification (n=149). In all models, age is included per year unit. ENT involvement was significantly associated with higher baseline eGFR. 95% CI, 95% confidence interval; ENT, ear-, nose-, and throat; eGFR, estimated glomerular filtration rate; PR3-ANCA, anti-proteinase 3 anti-neutrophil cytoplasm antibody; IFTA, interstitial fibrosis and tubular atrophy; AAGN, anti-neutrophil cytoplasm antibody associated glomerulonephritis.

Supplementary table 6. Multivariable regression analyses investigating the relationship between ENT involvement and eGFR in GPA patients

	β (95% CI)	P Value
ENT involvement	12.44 (0.08 – 24.80)	0.04
Age	-1.26 (-8.28 – 5.75)	0.02
PR3-ANCA	-5.17 (-17.02 – 6.68)	0.39
Tubulitis	-9.47 (-23.10 – 4.15)	0.17
Interstitial infiltrate	-16.29 (-24.93 – -7.64)	< 0.001
IFTA	-9.35 (-18.46 – -0.24)	0.01
AAGN classification	-9.79 (-15.58 – -4.00)	0.001

ENT involvement is associated with higher baseline eGFR in GPA patients. Age is included per year unit in the model. 95% CI, 95% confidence interval; ENT, ear-, nose-, and throat; eGFR, estimated glomerular filtration rate; PR3-ANCA, anti-proteinase 3 anti-neutrophil cytoplasm antibody; IFTA, interstitial fibrosis and tubular atrophy; AAGN, anti-neutrophil cytoplasm antibody associated glomerulonephritis.

Supplementary table 7. Multivariable regression analyses investigating the relationship between ENT involvement and 5-year follow-up eGFR

	Model investigating 5-year follow-up eGFR		Model investigating 5-year follow-up eGFR in PR3-ANCA positive patients	
	β (95% CI)	P Value	β (95% CI)	P Value
ENT involvement	2.12 (-4.90 – 9.14)	0.55	8.10 (-1.73 – 17.93)	0.10
Age	-0.27 (-0.52 – -0.03)	0.03	-0.03 (-0.37 – 0.30)	0.84
PR3-ANCA	-6.28 (-13.33 – 0.77)	0.08	N/A	N/A
Baseline eGFR	0.33 (0.17 – 0.48)	< 0.001	0.43 (0.24 – 0.61)	< 0.001
Tubulitis	4.25 (-4.43 – 12.92)	0.33	-1.92 (-13.49 – 9.64)	0.74
Interstitial infiltrate	-2.42 (-8.13 – 3.29)	0.40	-0.86 (-9.05 – 7.32)	0.83
IFTA	-1.65 (-8.23 – 4.92)	0.62	-2.73 (-11.14 – 5.69)	0.52
AAGN classification	-5.44 (-9.71 – -1.17)	0.01	N/A	N/A

ENT involvement is no longer associated with 5-year follow-up eGFR when baseline eGFR is included in the model. Both models are adjusted for within-trial therapy. Age is included per year unit in both models. 95% CI, 95% confidence interval; ENT, ear-, nose-, and throat; eGFR, estimated glomerular filtration rate; PR3-ANCA, anti-proteinase 3 anti-neutrophil cytoplasm antibody; IFTA, interstitial fibrosis and tubular atrophy; AAGN, anti-neutrophil cytoplasm antibody associated glomerulonephritis; N/A, not applicable.

Supplementary table 8. Three multivariable regression models investigating the association between baseline eGFR and other early disease manifestations

Model 1	Cutaneous model			Arthralgia/arthritis model			Lung model		
	β (95% CI)	P Value		β (95% CI)	P Value		β (95% CI)	P Value	
Cutaneous involvement	5.61 (-0.27 – 11.49)	0.06	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Arthralgia/arthritis	N/A	N/A	3.40 (-1.67 – 8.48)	0.19	N/A	N/A	N/A	N/A	N/A
Lung involvement	N/A	N/A	N/A	N/A	N/A	-2.63 (-7.56 – 2.30)	0.30		
Age	-0.66 (-0.84 – -0.48)	< 0.001	-0.63 (-0.81 – -0.45)	< 0.001	-0.69 (-0.87 – -0.51)	< 0.001			
PR3-ANCA	8.78 (3.74 – 13.82)	0.001	8.73 (3.65 – 13.80)	0.001	9.58 (4.56 – 14.60)	< 0.001			

Model 2	Cutaneous model			Arthralgia/arthritis model			Lung model		
	β (95% CI)	P Value		β (95% CI)	P Value		β (95% CI)	P Value	
Cutaneous involvement	-0.76 (-8.47 – 6.95)	0.85	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Arthralgia/arthritis	N/A	N/A	-1.74 (-7.94 – 4.46)	0.58	N/A	N/A	N/A	N/A	N/A
Lung involvement	N/A	N/A	N/A	N/A	N/A	-5.46 (-11.46 – 0.53)	0.07		
Age	-0.48 (-0.71 – -0.25)	< 0.001	-0.40 (-0.63 – -0.16)	0.001	-0.48 (-0.71 – -0.25)	< 0.001			
PR3-ANCA	1.45 (-4.81 – 7.70)	0.65	0.67 (-5.65 – 6.99)	0.83	2.57 (-3.74 – 8.89)	0.42			
Tubulitis	-13.12 (-20.27 – -5.97)	< 0.001	-11.02 (-18.30 – -3.74)	0.003	-13.20 (-20.25 – -6.14)	< 0.001			
Interstitial infiltrate	-8.86 (-13.06 – -4.66)	< 0.001	-9.21 (-13.46 – -4.95)	< 0.001	-8.75 (-12.91 – -4.59)	< 0.001			
IFTA	-11.97 (-17.11 – -6.84)	< 0.001	-11.89 (-17.16 – -6.62)	< 0.001	-12.07 (-17.16 – -6.98)	< 0.001			

Model 3	Cutaneous model			Arthralgia/arthritis model			Lung model		
	β (95% CI)	P Value		β (95% CI)	P Value		β (95% CI)	P Value	
Cutaneous involvement	-2.98 (-11.58 – 5.62)	0.49	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Arthralgia/arthritis	N/A	N/A	-4.85 (-11.40 – 1.71)	0.15	N/A	N/A	N/A	N/A	N/A
Lung involvement	N/A	N/A	N/A	N/A	N/A	-4.74 (-11.19 – 1.70)	0.15		
Age	-0.63 (-0.87 – -0.38)	< 0.001	-0.54 (-0.78 – -0.29)	< 0.001	-0.61 (-0.85 – -0.37)	< 0.001			
PR3-ANCA	1.29 (-5.62 – 8.21)	0.71	0.41 (-6.52 – 7.33)	0.91	1.86 (-5.06 – 8.78)	0.60			

Supplementary table 8. Three multivariable regression models investigating the association between baseline eGFR and other early disease manifestations (Continued)

Model 3	Cutaneous model		Arthralgia/arthritis model		Lung model	
	β (95% CI)	P Value	β (95% CI)	P Value	β (95% CI)	P Value
Tubulitis	-16.54 (-24.62 – -8.47)	<0.001	-14.71 (-22.91 – -6.50)	0.01	-16.83 (-24.87 – -8.79)	<0.001
Interstitial infiltrate	-7.48 (-11.96 – -3.00)	0.001	-7.38 (-11.83 – -2.93)	0.001	-7.12 (-11.53 – -2.71)	0.002
IFTA	-9.57 (-15.62 – -3.51)	0.002	-9.92 (-16.01 – -3.83)	0.002	-9.97 (-15.97 – -3.97)	0.001
AAGN classification	-6.82 (-10.61 – -3.03)	0.001	-6.98 (-10.79 – -3.17)	<0.001	-6.79 (-10.49 – -3.09)	<0.001

Since renal biopsies were not available for all patients, including the histopathological parameters limited the number of patients included in the analysis. To be able to include all patients we therefore created three models. The first model included only clinical parameters (n=412). In the second model, we added tubulointerstitial parameters (n=195). The third model, we added the histopathological classification (n=149). In all models, age is included per year unit. 95% CI, 95% confidence interval; eGFR, estimated glomerular filtration rate; PR3-ANCA, anti-proteinase 3 anti-neutrophil cytoplasm antibody; IFTA, interstitial fibrosis and tubular atrophy; AAGN, anti-neutrophil cytoplasm antibody associated glomerulonephritis.

Supplementary table 9. Multivariable regression analyses investigating the relationships between 5-year follow-up eGFR and other early disease manifestations

	Cutaneous model		Arthralgia/arthritits model		Lung model	
	β (95% CI)	P Value	β (95% CI)	P Value	β (95% CI)	P Value
Cutaneous involvement	0.78 (-8.80 – 10.35)	0.87	N/A	N/A	N/A	N/A
Arthralgia/arthritits	N/A	N/A	0.00 (-8.62 – 8.62)	0.99	N/A	N/A
Lung involvement	N/A	N/A	N/A	N/A	6.99 (-1.19 – 15.17)	0.09
Age	-0.37 (-0.67 – -0.07)	0.02	-0.32 (-0.63 – -0.01)	0.04	-0.32 (-0.61 – -0.03)	0.03
PR3-ANCA	-4.19 (-12.48 – -4.09)	0.32	-5.39 (-13.67 – -2.89)	0.20	-5.66 (-13.97 – -2.65)	0.18
Tubulitis	-3.68 (-12.87 – 5.50)	0.43	-0.96 (-10.45 – 8.53)	0.84	-2.70 (-11.76 – 6.36)	0.56
Interstitial infiltrate	-4.84 (-11.66 – 1.99)	0.16	-6.89 (-13.77 – -0.02)	0.05	-4.74 (-11.42 – 1.94)	0.16
IFTA	-10.07 (-16.84 – -3.29)	0.004	-10.74 (-17.91 – -3.57)	0.004	-10.12 (-16.75 – -3.48)	0.003

To investigate whether our associations are specific to ENT involvement, analyses between eGFR at 5-year follow-up and other early disease manifestations were performed. There are no associations between eGFR at 5-year follow-up and cutaneous involvement, arthralgia/arthritits, or lung involvement. All models are adjusted for within-trial therapy. In all models, age is included per year unit. 95% CI, 95% confidence interval; eGFR, estimated glomerular filtration rate; PR3-ANCA, anti-proteinase 3 anti-neutrophil cytoplasm antibody; IFTA, interstitial fibrosis and tubular atrophy; AAGN, anti-neutrophil cytoplasm antibody associated glomerulonephritis; N/A, not applicable

III. Supplementary data Chapter VI

Effect of rituximab on malignancy risk in patients with ANCA-associated vasculitis

<i>Table S1. Subgroup analysis</i>	<i>312</i>
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Supplementary table 1. Subgroup analysis

	N patients	N observed malignancies	SIR (95% CI)*	SIR p value*	RR (95% CI)*	RR p value*
Gender						
Male	149	20	1.58 (0.96–2.44)	0.07	1 (reference)	
Female	174	25	2.25 (1.45–3.32)	<0.001	1.43 (0.76–2.71)	0.30
Age at diagnosis						
≥59 years	159	29	1.60 (1.07–2.29)	0.02	1 (reference)	
<59 years	164	16	2.84 (1.62–4.61)	<0.001	1.78 (0.90–3.38)	0.10
Clinical diagnosis						
Microscopic polyangiitis	160	23	1.59 (1.01–2.38)	0.05	1 (reference)	
Granulomatosis with polyangiitis	109	14	2.20 (1.20–3.68)	0.01	1.39 (0.66–2.81)	0.43
Eosinophilic granulomatosis with polyangiitis	54	8	2.75 (1.19–5.41)	0.02	1.73 (0.67–4.02)	0.27
ANCA serotype†						
MPO-ANCA	110	15	1.56 (0.87–2.58)	0.13	1 (reference)	
PR3-ANCA	152	24	2.18 (1.40–3.25)	<0.001	1.40 (0.70–2.87)	0.39
Renal transplantation						
No	311	41	1.79 (1.29–2.43)	<0.001	1 (reference)	
Yes	12	4	4.31 (1.17–11.04)	0.03	2.40 (0.62–6.62)	0.20
Follow-up						
0–5 years	156	16	2.38 (1.36–3.86)	<0.001	1 (reference)	
5–10 years	135	23	1.81 (1.15–2.72)	0.01	0.76 (0.39–1.54)	0.50
>10 years	32	6	1.38 (0.51–3.00)	0.55	0.58 (0.19–1.56)	0.35

* The standard incidence ratio (SIR) is the ratio of observed to expected malignancies and represents the malignancy risk compared to the general population, and the relative risk (RR) represents the malignancy risk compared to the reference group. Calculated by exact Poisson regression analysis.

† Unknown for 61 patients.

MPO-ANCA, myeloperoxidase ANCA; PR3-ANCA, proteinase 3 ANCA.

Supplementary table 2. Cyclophosphamide and rituximab treatment in the subgroups

	N patients treated with cyclophosphamide	Duration of cyclophosphamide treatment, months (SD)	Mean cumulative cyclophosphamide dose, g (SD)	N patients treated with rituximab	Duration of rituximab treatment, months (SD)	Mean cumulative rituximab dose, g (SD)
Gender						
Male	107	5.5 (5.5)	9.9 (6.5)	68	22.1 (14.9)	5.8 (3.4)
Female	116	6.4 (14.4)	8.4 (10.8)	85	20.9 (14.4)	5.9 (3.4)
Age at diagnosis						
≥59 years	114	6.5 (13.8)	6.8 (3.9)	63	20.2 (14.2)	5.2 (2.9)
<59 years	109	5.1 (4.2)	11.5 (11.8)	90	22.5 (14.9)	6.3 (3.6)
Clinical diagnosis						
Microscopic polyangiitis	116	6.3 (12.6)	8.1 (10.5)	65	18.7 (13.3)	5.2 (2.6)
Granulomatosis with polyangiitis	88	5.4 (4.4)	10.7 (7.3)	66	27.7 (15.0)	6.7 (4.0)
Eosinophilic granulomatosis with polyangiitis	19	4.5 (2.8)	7.8 (4.2)	22	22.6 (15.7)	5.3 (2.9)
ANCA serotype*						
MPO-ANCA	72	7.4 (16.1)	7.2 (4.7)	43	20.7 (16.9)	5.1 (2.9)
PR3-ANCA	121	5.0 (4.2)	9.2 (6.5)	82	20.9 (13.0)	6.1 (3.3)
Renal transplantation						
No	213	5.2 (4.9)	9.1 (9.0)	149	21.6 (14.4)	5.9 (3.4)
Yes	10	15.6 (37.3)	8.5 (8.6)	4	16.7 (19.4)	4.5 (2.4)
Follow-up						
0–5 years	102	4.7 (3.5)	7.0 (4.6)	52	17.5 (10.9)	4.6 (2.8)
5–10 years	101	7.2 (15.5)	10.8 (11.9)	82	23.5 (15.2)	6.3 (3.6)
>10 years	20	5.4 (5.1)	10.9 (6.8)	19	24.6 (19.6)	7.4 (3.1)

* Unknown for 61 patients.

MPO-ANCA, myeloperoxidase ANCA; PR3-ANCA, proteinase 3 ANCA.

Supplementary table 3. SIR for non-melanoma skin cancer according to cumulative cyclophosphamide and rituximab dose*

Cumulative dose (g)	N patients	N observed non-melanoma skin cancer	SIR (95% CI)†	SIR p value†
Cyclophosphamide				
0	89	3	2.17 (0.45– 6.34)	0.16
0.1–20	207	18	4.89 (2.90 – 7.72)	<0.001
20–108	16	3	11.72 (2.42 – 34.25)	0.002
Rituximab				
0	167	23	8.47 (5.37 – 12.71)	<0.001
0.1–6	70	1	0.83 (0.02 – 4.64)	0.66
6–18	83	0	0 (0 – 2.47)	0.23

* SIR, standardised incidence ratio; the SIR is the ratio of the observed to expected malignancies adjusted for sex, age (per 5-year age group), and calendar time period (per 1-year calendar time period).

† Calculated by exact Poisson regression analysis.

Supplementary table 4. Cumulative cyclophosphamide and rituximab dose of each patient with a malignancy

Malignancy or malignancy site	N observed malignancies	Cumulative cyclophosphamide dose (g)*	Cumulative rituximab dose (g)*	Time to malignancy (years)†
Lung	4	36.0; 21.1; 4.9; 0.0	0.0; 7.0; 0.0; 5.0	3.7; 4.8; 0.5; 8.5
Breast	3	18.0; 6.0; 3.4	0; 4.0; 8.0	0.9; 1.1; 4.0
Colon or rectum	3	13.8; 9.5; 4.0	0.0; 0.0; 0.0	2.4; 4.5; 4.0
Prostate	2	10.0; 0.0	0.0; 0.0	7.4; 1.2
Bladder	1	0.0	1.0	2.4
Pancreas	1	0.0	0.0	1.4
Testis	1	7.0	5.0	4.6
Ovary	1	3.0	6.6	2.4
Melanoma	1	3.3	0.0	1.8
Tongue	1	0.0	0.0	4.5
Central nervous system	1	2.0	5.6	3.2
Kidney	1	7.0	4.0	2.4

* The cumulative doses of each patient with a malignancy is given. Patients with a cumulative dose of 0.0 did not receive the treatment. When more cases of the malignancy were observed, the first reported cumulative cyclophosphamide dose corresponds to the first reported cumulative rituximab dose, and to the first reported time to malignancy.

† This is the time between the date of diagnosis of ANCA-associated vasculitis and the date of diagnosis of the malignancy.

