

Supplementary information S1 (Table) | **Genes located on the human X chromosome with a direct or indirect role in immune functions***

Gene Location	Gene	Description	Human map	Function in immunity and/ or disease	Mouse map
Pseudoautosomal region 1 (PAR1)	<i>CRFL2</i>	Cytokine receptor-like factor 2	Xp22.3; Yp11.3	Enhances DC maturation and T cell proliferation	Chr. 5
	<i>GMCSFR/CSF2RA</i>	Granulocyte/macrophage colony-stimulating factor	Xp22.3; Yp11.3	Granulocyte and monocyte differentiation	Chr. 19
	<i>IL3RA</i>	Interleukin 3 receptor alpha	Xp22.3; Yp11.3	Component of the IL-3 receptor	Chr. 14
	<i>CD99/MIC2</i>	CD99 antigen	Xp22.3; Yp11.3	T cell regulation	Chr. 4
Nonrecombining region of the X chromosome (short arm)	<i>ARHGAP6</i>	Rho GTPase activating protein 6	Xp22.2	Regulation of the small GTP-binding proteins of the RAS superfamily	X (F5)
	<i>TLR7</i>	Toll-like receptor 7	Xp22.2	Viral recognition; SLE	X (F5)
	<i>TLR8</i>	Toll-like receptor 8	Xp22.2	Viral recognition; susceptibility to tuberculosis	X (F5)
	<i>PIGA</i>	Phosphatidylinositol glycan anchor biosynthesis class A	Xp22.2	PNH; regulation of GPI-anchored complement regulatory proteins	X (F3-F4)
	<i>BMX</i>	Bone marrow kinase X-linked	Xp22.2	Growth and differentiation of haematopoietic cells	X (F)
	<i>RPS6KA3/RSK2</i>	Ribosomal protein S6 kinase alpha-3	Xp22.1	Control of TLR signalling; required for endocytosis in DCs	X (F4)
	<i>CNKSR2</i>	Connector enhancer of kinase suppressor of ras 2	Xp22.1	Regulator of RAS signalling	X (F4)
	<i>IL1RAPL1</i>	IL1 receptor associated protein-like 1	Xp21.3	Orphan receptor of the IL-1R superfamily	X (C1)
	<i>CYBB/NOX2</i>	Cytochrome b-245 beta polypeptide	Xp11.4	X-CGD; ROS production	X (A1.1)
	<i>DUSP21</i>	Dual specificity phosphatase 21	Xp11.3	Phosphatase activity	X (A1.2)
	<i>TIMP1</i>	Tissue inhibitor of metalloproteinase 1	Xp11.23	RA; CD; inhibitor of metalloproteinases; erythroid-potentiating activity; wound repair	X (A1.3)
	<i>PFC/CFP</i>	Properdin P factor complement	Xp11.23	Properdin deficiency; positive regulator of the alternative complement pathway	X (A3)
	<i>WAS</i>	Wiskott-Aldrich syndrome protein	Xp11.23	WAS; cytoskeleton organization	X (A1.1)
	<i>GATA1</i>	GATA binding protein 1	Xp11.23	Differentiation of erythrocytes and megakaryocytes	X (A2)
	<i>FOXP3</i>	Forkhead box P3	Xp11.23	IPEX syndrome; differentiation of regulatory T cells	X (A1.1)
Nonrecombining region of the X chromosome (long arm)	<i>ARHGEF9</i>	CDC42 guanine nucleotide exchange factor (GEF) 9	Xq11.2	Activation of CDC42	X (C3)
	<i>VSIG4</i>	V-set and immunoglobulin domain containing 4	Xq12	Macrophage mediated phagocytosis; inhibition of T cell activation	X (C3)
	<i>EDA2R/XEDAR</i>	Ectodysplasin A2 receptor	Xq12	Activation of NF-κB and JNK pathways	X (C3)
	<i>EDA</i>	Ectodysplasin A	Xq13.1	XHED; regulation of NF-κB and JNK pathways	X (C3)
	<i>IGBP1</i>	Immunoglobulin (CD79A) binding protein 1	Xq13.1	Inhibitor of apoptosis	X (C3)
	<i>IL2RG</i>	Interleukin 2 receptor gamma	Xq13.1	X-SCID; receptor for interleukins 2, 4, 7, 9, 15 and 21	X (D)
	<i>CXCR3</i>	Chemokine (C-X-C motif) receptor 3	Xq13.1	Receptor for CXCL9, 10 and 11	X (D)
	<i>FGF16</i>	Fibroblast growth factor 16	Xq21.1	Cell growth, morphogenesis, tissue repair, inflammation	X (C3)
	<i>NOX1</i>	NADPH oxidase 1	Xq22.1	ROS production	X (E3)
	<i>BTK</i>	Bruton agammaglobulinaemia tyrosine kinase	Xq22.1	XLA; TLR4 and TLR2 signalling pathways	X (E3)

SUPPLEMENTARY INFORMATION

Nonrecombining region of the X chromosome (long arm)	<i>IL1RAPL2</i>	IL1 receptor-associated protein-like 2	Xq22.3	Orphan receptor of the IL1R superfamily	X (F1)
	<i>TSC22D3/GILZ</i>	TSC22 domain family member 3	Xq22.3	Anti-inflammatory and immunosuppressive functions	X (F1)
	<i>IL13RA2</i>	Interleukin 13 receptor alpha 2	Xq23	IL-13 signalling (decoy receptor)	X (F2)
	<i>IL13RA1</i>	Interleukin 13 receptor alpha 1	Xq24	IL-13 signalling	X (A3.3)
	<i>NKRF</i>	NF-kappaB repressing factor	Xq24	Silencing of IFNB through NF-kB inhibition	X (A3.3)
	<i>NKAP</i>	NF-kappaB activating protein	Xq24	Regulation of NF-kB activation; T cell development	X (A3.3)
	<i>XIAP/BIRC4</i>	Inhibitor of apoptosis X-linked	Xq25	XLP2; direct inhibition of caspases 3 and 7	X (A3-A5)
	<i>SH2D1A/SAP</i>	SLAM-associated protein	Xq25	XLP1/EBV infection	X (A5)
	<i>AIFM1</i>	Apoptosis-inducing factor	Xq26.1	Inducer of apoptosis	X (A6)
	<i>CD40L</i>	CD40 ligand	Xq26.3	XHIM syndrome; SLE; Malaria; HIV	X (A5)
	<i>ARHGEF6</i>	RAC/CDC42 guanine nucleotide exchange factor (GEF) 6	Xq26.3	Activation of the RAS-like family of Rho proteins; cytoskeleton organization; apoptosis	X (A5)
	<i>BGN</i>	Biglycan precursor	Xq28	Endogenous ligand for TLR2 and TLR4 in macrophages; activation of p38, ERK and NF-kB	X (B)
	<i>DUSP9/MKP4</i>	Dual specificity phosphatase 9	Xq28	Inactivation of MAP kinases	X (B)
	<i>ARHGAP4</i>	Rho GTPase activating protein 4	Xq28	Regulation of the small GTP-binding proteins of the RAS superfamily	X (A7.3)
	<i>IRAK1</i>	Interleukin-1 receptor-associated kinase 1	Xq28	TLR signalling	X (A7.3)
	<i>G6PD</i>	Glucose-6-phosphate dehydrogenase	Xq28	G6PD-deficiency; required for oxidative burst by phagocytes and erythrocytes	X (A7)
	<i>IKBKG/NEMO</i>	Inhibitor of kappaB kinase gamma	Xq28	Incontinentia pigmenti; EDA-ID; regulation of NF-kB signalling; susceptibility to mycobacterial diseases	X (A7.3)
<i>GAB3</i>	GRB2-associated binding protein	Xq28	Macrophage differentiation	X (A7.3)	
<i>MTCP1</i>	Mature T cell proliferation 1	Xq28	T cell proliferation	X (A7.3)	
Pseudoautosomal region 2 (PAR2)	<i>IL9R</i>	Interleukin-9 receptor	Xq28	IL-9 receptor on several haematopoietic cells; RA	Chr. 11

CD: Crohn's disease; CXCL: chemokine CXC motif; DC: dendritic cells; EBV: Epstein-Barr virus; EDA-ID: anhidrotic ectodermal dysplasia with immunodeficiency; ERK: extracellular signal-regulated kinases; GPI: glycosylphosphatidylinositol (anchor); GRB: growth factor receptor-bound protein; GTP: guanosine triphosphate; HIV: human immunodeficiency virus; IFN: interferon; IL: interleukin; IPEX: immune dysregulation-polyendocrinopathy-enteropathy, X-linked; JNK: c-Jun N-terminal kinases; MAP: mitogen-activated protein; NADPH: reduced form of nicotinamide adenine dinucleotide phosphate; NF-kB: nuclear factor kappa-B; p38: p38 MAP kinase; PNH: paroxysmal nocturnal hemoglobinuria; RA: rheumatoid arthritis; RAC: RAS-related C3 botulinum toxin substrate; RAS: rat sarcoma; ROS: reactive oxygen species; SLAM: signaling lymphocyte activation molecule; SLE: systemic lupus erythematosus; TLR: toll-like receptor; TSC22: transforming growth factor-beta-stimulated clone-22; WAS: Wiskott-Aldrich syndrome; X-CGD: chronic granulomatous disease, X-linked; XHED: X-linked hypohidrotic ectodermal dysplasia; XHIM: X-linked hyper-IgM syndrome; XLA: Bruton (X-linked) agammaglobulinemia; XLP: X-linked lymphoproliferative disease; X-SCID: severe combined immunodeficiency, X-linked. * The table depicts the genes located in the pseudoautosomal and nonrecombining regions of the human X chromosomes with direct or indirect immune functions. The depicted genes located in the nonrecombining region of the X chromosome do not have any known Y homologues. Mapping positions (chromosome number; cytogenetic band in brackets) of the mouse genome are shown for comparison. Human and mouse mapping positions are based on the Ensembl database.