

Category	Name	B cells	T cells	Ig	Inheritance	Notes
Lymphocyte Maturation	Btk Deficiency	decrease	normal	decrease	x-linked	B cell tyrosine kinase mutation
	X-Linked SCID	normal	decrease	decrease	x-linked	50% SCID, gamma-chain IL-receptor mutation, carrier females unaffected
	JAK3 Kinase SCID	normal	decrease	decrease	auto recess	receptor signaling kinase
	Adenosine DeAminase SCID	decrease	decrease	decrease	auto recess	purine salvage path - dATP buildup
	PNP SCID	normal	decrease	normal	auto recess	purine catabolism - dGTP buildup
	Reticular Dysgenesis	decrease	decrease	decrease	auto recess	defect hematopoietic stem cells
	RAG-1/2 Mutations	decrease	decrease	decrease	auto recess	defect antigen receptor recombination
	DiGeorge Syndrome	normal	decrease	normal	dom/hapinsf	failed 3+4th pharyngeal (thymus)
Lymphocyte Activation	IgA Deficiency	normal	normal	decr (A)	auto recess	50% silent
	IgG Deficiency	normal	normal	decr (G)	unknown	defect B cell differentiation
	X-Linked Hyper-IgM Syndrome	normal	normal	incr (M)	x-linked	T cells lack CD40L: no isotype switch
B Cell Differentiation	Common B Cell Diff. Disorder	norm/decr	normal	d (G,M,A)	sporadic	heterogenous disorder category
	X-Linked Lymphoproliferative Dis.	increase	increase	increase	x-linked	def. SAP adaptor protein, abnormal EBV, uncontrolled B cell and CD8T and hemophagocytosis
	Familial Hemophagocytic Syn.					inappropriate and ineffective T cells
Multi-System Diseases	Wiskott-Aldrich Syndrome	normal	decrease	decrease	x-linked	WASP protein, thrombocytopenia, excema, malignant
	Ataxia Telangiectasia	normal	decrease	dec (A,G)	auto recess	defect in DNA repair
	APECED					AIRE mutation, loss of central tolerance
	ALPS					FAS / FASL mutation
	IPEX		decr. CD4			FoxP3: loss of CD4 cells
Innate Immunity	Phagocytic Disorders					neutropenia, ox/non-ox metab. disorder
	Chronic Granulomatous Disease					defective oxidant superoxides, treat with INF-gamma
	Leukocyte Adhesion Def. Type 1				auto recess	loss B2 integrins and CD18, effects all imm. cells
	Chediak-Higashi Syndrome				auto recess	def. neut/mono/NK, albinism
	Complement Deficiencies (var)					def. inflam, opson, lysis, imm interact
HIV	HIV-Induced Immunodeficiency		decr. CD4			