Supplementary table 1 **| PADs related to B cell-intrinsic defects**

B cell-intrinsic defects

PAD	Affected protein	Inheritance	Affected cell lineage	Other manifestations
I-B cell development defects				
agammaglobulinemia	λ5, Cμ, Igα, Igβ, Pi3K (p85α), BLNK	AR	B	
	BTK	XL	B	
SCID	ADA, RAG	AR	B+T	
	AK2	AR	B+T+ PMN	deafness
	components of NHEJ	AR	B+T	
Dyskeratosis congenita (Hoyeraal–Hreidarsson)	Telomerase or shelterin complex	AR or XL	B+T	multi-organ defects (cerebellum defects)
'MonoMAC'/DCML deficiency	GATA2	AR	B+T+monocytes+ dendritic cells+NK cells	
II-B cell migration defects				
WHIM syndrome	CXCR4 (gain of function)	AD	B+PMN cells	
Wiskott Aldrich syndrome WAS-like	WASP	XL	B+T$^+$ platelets	
	WIP	AR	B+T$^+$ platelets	
Combined immunodeficiency	DOCK8	AR	B+T	
	MST1	AR	B+T	
III-B cell survival				
Common variable immunodeficiency	BAFFR/TACI	AR/AR or AD	B	
	TWEAK	AD	B (DN T, NK?)	
IV- B cell activation defects				
ICF syndrome	DNMT3B, ZBTB24	AR	B+T	multi-organ defects
hypogammaglobulinaemia	CD19 complex, CD20	AR	B	
combined immunodeficiency	CARD11, IκBβ	AR	B+T	
	CARD11 (gain of function)	AD	B	
Selective polysaccharide antibody deficiency	NEMO	XL	B+T	multi-organ defects
	IκBα (gain of function)	AD	B+T	multi-organ defects
	HOIL1	AR	B	auto-inflammation
	ORAI1, STIM1	AR	B+T	Muscle defects
	LRBA	AR	B+T	
	POLε	AR	B+T	Bone defects
PLCγ2associated antibody deficiency and immune dysregulation	PLCγ2(gain of function)	AD	B	allergy, auto-immunity
V-CSRD				
	CD40	AR	B+T+monocytes	
	AID	AR (AD)	B	
	UNG	AR	B	
AT, NBS	PMS2	AR	B+T	multi-organ defects
	ATM, MRE11, NBS1, RNF168	AR	B+T	multi-organ defects
SCID	Components of NHEJ	AR	B+T	

VI- Defective response to cytokines				
combined immunodeficiency	IL21R	AR	B+T	
Hyper-IgE syndrome	STAT3	AD	B+T	Bone and blood vessel defects
Severe combined immunodeficiency	γc	XL	T+NK	
	Jak 3	AR	T+NK	

AD: autosomal dominant, AR: autosomal recessive, AT: Ataxia-Telangiectasia, CSRD: class switch recombination deficiencies, CVID: common variable immunodeficiency, DCML-deficiency: dendritic cell, monocyte, B and NK lymphoid deficiency, DN T: double negative T cells, FILS: facial dysmorphism, immunodeficiency, livedo and short stature, HL: haematopoietic lineage, ICF: immunodeficiency, centromeric instability, facial anomalies, MonoMAC: monocytopenia, mycobacterial infections, NBS: Nijmegen breakage syndrome, NHEJ: Non homologous end joining, PMN: polymorphonuclear cells, SCID: severe combined immunodeficiency, WHIM: wharts, hypogammaglobulinaemia, infections and myelokathexis, XL: chromosome X-linked.