

Supplementary table 2 | **PADs related to B cell-extrinsic defects**

PAD	Affected protein	Inheritance	Affected cell lineage	Other manifestations
I-Defective T cell differentiation				
Combined immunodeficiency	CD3 subunits, IL7R α	AR	T	
Di George syndrome	22q11.2deletion	AD	T	multi-organ defects
II- Defective T cell function				
Combined immunodeficiency	ZAP70, ITK, p56LCK	AR	T	
	Regulatory factors of MHC class II transcription	AR	T+B+monocytes+dendritic cells	
III- Defective T_{HH}				
X lymphoproliferative and related disease	SAP	XL	T+B	
	CD27	AR	T+B+NK	
Hyper-IgE syndrome	STAT3	AD	T+B	Bone and blood vessel defects
CSRD	CD40L	XL	T	
CVID	ICOS	AR	T	
IV- Innate immunity defect				
TLR/IL1R signaling defect	IRAK4, MYD88, TIRAP	AR	T+B ⁺ monocytes + dendritic cells	multi-organ defects

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.