

Supplementary Table 3: Frequency of patients with inborn errors of immunity showing defects of the B-cells subsets identified in the EF PIDOT, as compared to age-reference values.

Disease	PreGC	MBC/PC	Unswitched MBC/PC	Switched MBC/PC	Any of them
SCID (n=24)	75%	100%	100%	100%	100%
IL2Rg	1/6	6/6	6/6	6/6	6/6
IL7R	0/1	1/1	1/1	1/1	1/1
RAG1	8/8	8/8	8/8	8/8	8/8
RAG2	5/5	5/5	5/5	5/5	5/5
DCLRE1C	3/3	3/3	3/3	3/3	3/3
NHEJ1	1/1	1/1	1/1	1/1	1/1
CID (n=12)	8%	42%	42%	83%	83%
CD40L	0/6	1/6	1/6	6/6	6/6
ZAP70	0/3	1/3	1/3	1/3	1/3
DOCK8	1/2	2/2	2/2	2/2	2/2
BCL10	0/1	1/1	1/1	1/1	1/1
CID with syndromic features (n=20)	25%	45%	45%	45%	60%
WASp	0/3	3/3	3/3	1/3	3/3
ATM	3/6	4/6	3/6	4/6	4/6
Di George	1/6	1/6	1/6	1/6	1/6
STAT3	0/2	0/2	1/2	1/2	2/2
NEMO	0/2	1/2	1/2	1/2	1/2
PNP	1/1	0/1	0/1	1/1	1/1
PAD (n=16)	75%	94%	94%	94%	100%
BTK	10/10	10/10	10/10	10/10	10/10
PI3KCD	2/5	5/5	5/5	4/5	5/5
AID	0/1	0/1	0/1	1/1	1/1
Disease of immune dysregulation (n=10)	20%	40%	40%	50%	60%
Syntaxin	1/1	1/1	0/1	1/1	1/1
FAS	0/5	2/5	2/5	2/5	2/5
XLP	0/1	0/1	0/1	1/1	1/1
CD27	0/1	0/1	1/1	0/1	1/1
CTPS1	1/2	1/2	1/2	1/2	1/2
Defects of phagocytes or function (n=10)	20%	40%	30%	50%	60%
CGD	0/5	2/5	2/5	2/5	3/5
GATA2	2/5	2/5	1/5	3/5	3/5
Defects innate immunity (n=3)	33%	67%	67%	67%	67%
STAT1	0/1	1/1	1/1	1/1	1/1
WHIM	1/1	1/1	1/1	1/1	1/1
IRAK4	0/1	0/1	0/1	0/1	0/1
Complement deficiencies (n=4)	0%	0%	0%	0%	0%

Results expressed as percentage of patients showing absolute counts below the lower limit of normality, compared to age-reference values obtained from 250 healthy donors analyzed with the same protocol. SCID: Severe Combined Immunodeficiency. CID: Combined Immunodeficiency. PAD: Predominantly Antibody Deficiency. MBC/PC: Memory B-cells/Plasma cells.