

Supplementary Table 1 Immunophenotyping, *in vitro* T-cell proliferation, serum immunoglobulin levels and vaccination titers.

(a) Immunophenotyping of peripheral lymphocytes. Reference values: Division of Rheumatology and Clinical Immunology, University Hospital Freiburg, Germany.

	Normal Range	#5	#10	#12	#13
Total Number of Lymphocytes	1500-4000	4216	6254	4290	6603
CD2⁺CD18⁺ Lymphocytes	1050-3680	3464	4472	3050	4654
CD3⁺ T-Cells	990-3360	3414	5586	2896	4549
TCRαβ⁺ of CD3⁺ T-Cells	1335-3920	4071	5402	4062	6078
TCRαβ⁺CD4⁺CD8⁺ of CD3⁺ T-Cells	8-100	33	43	30	52
TCRαβ⁺CD4⁺CD8⁻ of CD3⁺ T-Cells	6-60	53	112	48	86
TCRγδ⁺ of CD3⁺ T-Cells	14-440	127	817	201	466
HLA-DR⁺ of CD3⁺ T-Cells	36-440	1766	881	668	683
CD28⁺ of CD3⁺ T-Cells	1215-3960	2943	4604	3730	5503
CD29⁺ of CD3⁺ T-Cells	345-2720	2601	3044	2230	2395
CD45RA⁺ of CD3⁺ T-Cells	735-2880	2858	4334	2546	4697
CD4⁺ T-Cells	600-2320	1036	2206	1364	2740
CD28⁺ of CD4⁺ T-Cells	1440-4000	3952	6079	4264	6411
CD29⁺ of CD4⁺ T-Cells	345-2480	1826	1788	1384	1510
CD45RA⁺ of CD4⁺ T-Cells	630-2720	2441	4185	2661	4649
CD8⁺ T-Cells	240-1240	2351	1817	1453	1622
CD11b⁺ of CD8⁺ T-Cells	60-1480	1334	3043	1508	2816
CD57⁺ of CD8⁺ T-Cells	27-1480	1122	2337	660	1916
CD38Fl⁺ of CD8⁺ T-Cells	120-720	801	1438	644	2575
CD5⁺ Lymphocytes	840-3480	3292	5333	2814	4374
CD3⁻CD16⁺ NK-Cells	90-1000	113	307	109	235
CD19⁺ B-Cells	90-600	715	1523	1266	1781
CD40⁺ of B-Cells (%)	>90	97.55	99.57	99.87	98.47
CD5⁺ of B-Cells (%)	20-47	13.72	27.79	25.91	26.42
IgD⁺IgM⁺CD27⁻ of B-Cells (%)	38-80	94.2	85.5	91.9	87.1
IgD⁺IgM⁺ of B-Cells (%)	66-87	97.6	94.3	97.1	95
IgD⁻IgM⁻ of B-Cells (%)	10-31	1.9	5.1	2.7	4.2
IgD⁺IgM⁺CD27⁺ Of B-Cells (%)	9-33	5.5	8.7	5.3	8
IgD⁻IgM⁻CD27⁺ of B-Cells (%)	9-30	1.6	4.7	2.1	3.4
IgA⁺ B-Cells (%)	3-13	0.8	1.5	1	0.6
CD21⁺ of B-Cells (%)	>88	93.1	92.7	95.9	84.9
CD40⁺ Of B-Cells	>1350	4113	6227	4284	6502
CD5⁺ Of B-Cells	300-1880	578	1738	1112	1745
IgM⁺IgD⁺ Of B-Cells	990-3480	4115	5898	4166	6273
IgM⁻IgD⁻ Of B-Cells	150-1240	80	319	116	277
IgD⁺CD27⁺ Of B-Cells	135-1320	232	544	227	528
IgD⁻CD27⁺ Of B-Cells	135-1200	67	294	90	225
IgA⁺ B-Cells	45-1200	34	94	43	40
CD21⁺ Of B-Cells	>1320	3925	5797	4114	5606

(b) *In vitro* T-cell proliferation upon indicated stimuli. cpm: counts per minute after ³H-TdR incorporation; PHA: phytohemagglutinine; ConA: concavalin A; PWM: pokeweed mitogen. Reference values: Division of Rheumatology and Clinical Immunology, University Hospital Freiburg, Germany.

	Normal Range	#5	#10	#12	#13
Medium (cpm)	400-2000	417	568	1271	976
PHA 1 µg/ml (cpm)	>40000	131640	96999	127102	122149
ConA 10 µg/ml (cpm)	>20000	100509	61387	79233	68764
Anti-CD3 10 ng/ml (cpm)	>34000	49092	45061	82157	69742
PWM 1:800 (cpm)	>10000	28025	29679	42760	37571
Tuberculin 25 µg/ml (cpm)	>1000	727	959	1713	1178
Tetanus Toxoid 10 µg/ml (cpm)	>1000	699	1073	1505	1533
Candida 0.5 µg/ml (cpm)	>1000	853	1186	1691	721
IL-2 20 IU/ml (cpm)	>1000	7996	7479	10303	5218
ConA+IL-2 (cpm)	>27000	95394	77637	101438	81427
Anti-CD3+IL-2 (cpm)	>34000	73171	58803	91319	63443

(c) Serum immunoglobulin levels and selected vaccination titers. Age adjusted reference values given in brackets. n.d.: not determined. Reference values: Department of Clinical Immunology, Hannover Medical School, Hannover, Germany.

	#5	#10	#12	#13
IgM (g/l)	0.21 (0.4-2.3)	0.2 (0.47-1.9)	0.28 (0.47-1.9)	0.34 (0.38-1.5)
IgA (g/l)	2.68 (0.7-4)	2.94 (0.47-2.1)	1.26 (0.47-2.1)	0.56 (0.27-1.5)
IgG (g/l)	6.17 (6.7-14)	7.32 (5.2-12.9)	5.55 (5.2-12.9)	4.02 (4.7-11.1)
IgG1 (g/l)	4.46 (4.9-11.4)	6.09 (2.7-12.9)	3.92 (2.8-17.4)	2.87 (3.5-7.9)
IgG2 (g/l)	1.33 (1.5-6.4)	1.28 (1.1-5.5)	1.19 (0.8-5.5)	0.77 (0.5-2.6)
IgG3 (g/l)	0.15 (0.2-1.1)	0.16 (0.13-2.5)	0.37 (0.2-3.2)	0.18 (0.1-2.3)
IgG4 (g/l)	0.16 (0.08-1.4)	0.13 (0.07-5.3)	< 0.02 (0.1-1.7)	<0.02 (0.05-1.8)
IgE (IE/ml)	2 (1-100)	6 (1-132)	8 (2-155)	5 (2-60)
Anti-Tetanus IgG (IU/ml)	<0.01 (>0.01)	<0.01 (>0.01)	<0.03 (>0.01)	0.26 (>0.01)
Anti-Diphtheria IgG (IU/ml)	<0.01 (>0.1)	<0.01 (>0.1)	<0.01 (0.1)	0.02 (>0.1)
Anti-HiB IgG (µg/ml)	1.13 (>1)	0.97 (>1)	n.d.	>9.5 (>1)
Anti-Pneumococcus Titer	1:1000 (1:8000)	1:1000 (1:8000)	n.d.	1:1000 (1:4245)