Table S1. Blood leukocyte subsets in IRAK-4–deficient patients

Patients (age years)	P11 1	P23 1	P19 2	Normal values 1–2	P1 5	P8 5	P15 5	P28 5	P3 7	Normal values 5–7	P2 14	P17 27	P18 27	P7 32	Normal values >14
Lymphocytes (10^9 per µl)	5	6.2		(3.6–9)	3.3	1.9	4.8	3.2	5	(2.3–5.7)	2.6	1.3	1.3		(1.4–3.3)
T cells (%)															
$CD3^+$	71	77	73	(53–75)	75	71	86	57	68	(56–76)	75	72	76		(56–84)
$CD4^+$	48	48	29	(32–51)	55	45	63	35	52	(28–47)	54	56	46		(31–52)
$CD8^+$	29	26	43	(14–30)	32	25	17	24	14	(16–35)	22	13	20		(18–35)
$CD4^+/CD45RA^+$		40		(64–93)							58				
$CD4^+/CD45RO^+$											45				
$CD8^+/CD45RA^+$											86				
NK cells (%)															
$CD56^+/CD16^-$		5		(4–31)			3.4	3	2	(4–31)	10				(4–31)
$CD56^+/CD3^-$						4	8.3			(4–22)	12	11	8		(4–22)
CD56 *bright*							3.4			(3–13)					
CD56 *dim*							96.6			(87–97)					
$CD56^+/CD3^+$							0.2			(2–8)					
B cells (%)															
$CD19^+$	7	10	17	(6–35)	23	20	6	38	28	(6–35)	16	15	12		(6–35)
$CD19^+/CD27^+$											17				(10–25)
Monocytes (%)															
$CD14^+/CD16^-$						6.8				(6.2–15.4)	4.6			12.4	(6.2–15.4)
$CD14^+/CD16^+$						0.4				(0.15–1.4)	0.33			0.1	(0.15–1.4)
CD16 high						0.7				(1.05–2.6)	0.83			1.72	(1.05–2.6)
BDCA-1^+						0.11				(0.09–0.9)	0.48			0.12	(0.09–0.9)
BDCA-2^+						0.31				(0.15–0.6)	0.8			0.35	(0.15–0.6)

Total lymphocyte counts; percentages of T cells, NK cells, B cells, and monocytes; and the age-specific normal values are shown.

Table S2. T cell proliferation, Ig levels, and humoral responses to recall antigens and to glycans in IRAK-4–deficient patients

Patients (age)	P6 (2 mo)	P5 (1 y)	P11 (1 y)	P23 (1 y)	Normal values (1–2 y)	P1 (5 y)	P8 (5 y)	P15 (5 y)	P13 (7 y)	P3 (7 y)	Normal values (5–7 y)	P2 (14 y)	P24 (16 y)	P17 (27 y)	P18 (27 y)	P7 (32 y)	Normal values >14 y
Proliferation ($\times 10^3$ cpm)																	
CD3				59	(>30)						(>30)	80		26	48		(>30)
PHA					(>50)			142			(>50)	238		112	135		(>50)
PPD					(>10)			7			(>10)	94		1.7	10		(>10)
Candidine					(>10)			7			(>10)	16		7	19		(>10)
Tetanus					(>10)						(>10)	123		0.5	1		(>10)
S. aureus				35	(>10)						(>10)						(>10)
Serum Ig (mg/ml)																	
IgG	5.7	5.4	17	8.9	(5–10)	11.7	13.9	10.7			(7–12)	10.9		15	13	16.7	(9–14.8)
IgG1							8.4	5.88			(>4)						
IgG2							3.12	1.18			(>0.4)						
IgG3							0.34	0.45			(>0.16)						
IgG4							3.54	0.5			–						
IgA	0.6	0.2	0.9	0.47	(0.2–0.8)	0.63	3.35	1.31			(0.7–1.6)	0.22		0.3	0.6	1.1	(1.1–2.6)
IgM	0.6	1.1	2	1.11	(0.5–1.1)	0.72	0.74	0.5			(0.5–1.2)	0.82		1.3	1.5	1.9	(0.88–1.8)
IgE (kU/ml)			2,600	801	(<40)	17,400	187	352	792		(<100)	11	36.6	255	96.5	136	(<114)
Specific antibodies																	
Antitetanus			>0.1	0.12	(>0.1)		0.59	0.06		1.81	(>0.1)	0.47		0.34	0.45	0.06	(>0.1 IU/ml)
Poliovirus					(>40)			>40			(>40)	40					(>40 Ab)
Diphtheria				0.26	(>0.1)	2.04	0.1	0.002		0.88	(>0.1)					0.18	(>0.1 IU/ml)
S. pneumoniae				<0.3	(>0.3)	1.9	<0.3	>0.6			(>0.3)	<0.3		<0.3	<0.3	>0.3	(>0.3 μg/ml)
H. influenzae					pos.	pos.	pos.			pos.	pos.	pos.				pos.	pos.
Allohemagglutinin			1/16			1/128	1/2	1/8	1/8		≤1/16		1/32	1/16	1/16	1/2	≤1/16

Proliferative responses to OKT3 (CD3), to the mitogen PHA, and to different antigens (PPD, candidine, tetanus, and heat-killed *S. aureus*), serum Ig levels, and titers for specific antibodies and the age-specific normal values are shown. pos., positive.

Table S3. Serology of patients to common viruses

Patients Serology	P1	P2	P3	P7	P8	P13	P15
CMV	negative	negative	negative	negative	positive	IgG positive	
EBV	positive	positive	negative	positive	negative	positive	
EA	negative	negative					
VCA	IgG positive	IgG positive	negative	IgG positive	negative	IgG positive	
EBNA	negative	IgG positive	negative	IgG positive	negative	IgG positive	
HSV	negative	negative	negative	negative	negative	negative	
VZV	IgG positive	IgG positive	negative	IgG positive	negative	IgG positive	
HHV6	IgG positive	IgG positive	IgG positive	IgG positive	IgG positive	IgG positive	
Parvovirus B19	IgG positive	negative	negative	IgG positive	negative	negative	
Rubeola	IgG positive	IgG positive	IgG positive	IgG positive	IgG positive	negative	
Mumps	positive	positive	positive	positive	positive	positive	positive
coxsackie B1	negative	negative	negative	negative	negative	negative	negative
coxsackie B2	negative	positive	negative	negative	negative	positive	positive
coxsackie B3	positive	positive	negative	positive	positive	positive	positive
coxsackie B4	positive	positive	negative	negative	positive	negative	positive
coxsackie B6	negative	negative	negative	negative	negative	negative	negative
RSV	positive	positive	positive	positive	positive	positive	
HMPV	positive	negative	positive	positive	positive	positive	

EA, early antigen; EBNA, EBV nuclear antigen; HHV6, human herpes virus 6; HMPV, human metapneumovirus; RSV, respiratory syncytial virus; VCA, virus capsular antigen; VZV, varicella zoster virus.