Material Supplementary

Table S1. Current and ever experienced atopic manifestation per pediatric patient with a primary immunodeficiency disease

Patient number	Sex	Primary immunodeficiency disease	Ever atopic dermatitis	Current atopic dermatitis	Food allergy	Ever asthma	Current asthma	Ever hayfever	Current hayfever
1	M	hyper IgM syndrome	no	no		not applicable[§]	not applicable[§]	no	yes
2	F	SPAD	yes	no	no	yes	no	no	yes
3	M	IgA subclass deficiency	yes	yes	no	no	no	no	no
4	F	SPAD	no	no	no	yes	no	no	yes
5	F	hypogammaglobulinemia (IgG) and IgA subclass deficiency	no	no	no	yes	yes	no	no
6	F	IgG2 en IgG3 subclass deficiency	no	no	no	no	no	no	no
7	M	congenital neutropenia	yes	yes	diagnosis by doctor based on a double-blind placebo-controlled food challenge – cow's milk	no	no	yes	no
8	M	congenital neutropenia	yes	no	no	yes	no	yes	no
9	M	hypogammaglobulinemia	no	no		yes	yes	yes	no
10	M	hypogammaglobulinemia	yes	no	no	yes	yes	no	yes
11	M	congenital neutropenia	no	no	no	no	yes	yes	yes
12	F	APDS		no	no	no	yes	no	no
13	F	hypogammaglobulinemia (IgG, IgA)	yes	no	no	yes	no	yes	yes
14	M	CVID	no	no	no	yes	yes	no	yes
15	F	hypogammaglobulinemia	yes	no		no	no	no	yes
16	F	hypogammaglobulinemia (IgG)	no	no		no	no	no	no
17	M	CMC	no	no	no	no	no	no	no
18	F	IgG and IgM subclass deficiency	yes	no	diagnosis by doctor based on complaints – peanut – nuts	yes	no	yes	no
19	M	CVID		no	no	no	no		no
20	M	CVID(Cantu syndrome)	yes	yes	diagnosis by doctor based on a double-blind placebo-controlled food challenge – banana	yes	yes	yes	yes
21	M	CVID	yes	no	diagnosis by doctor based on complaints – egg white – milk		yes	yes	yes
22	M	hypogammaglobulinemia (IgG)	yes	yes		yes	no	no	
23	M	CVID	yes	no		no	no	no	no
24	F	agammaglobulinemia	no	no		not applicable[§]	not applicable[§]	no	no
25	M	agammaglobulinemia	yes	no		no	no	no	no
26	M	leukopenia (cartilage hair hypoplasia)	no	no		no	yes	no	no
27	M	XLA	yes	no		no	yes	no	yes

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28	M	interferon gamma receptor 1 deficiency	no	no	no	no	yes	no	no
29	M	hyper IgD syndrome	yes	no	diagnosis by doctor based on a double-blind placebo-controlled food challenge – cow's milk	not applicable[§]	not applicable[§]	no	no
30	M	hypogammaglobulinemia	yes	yes	diagnosis by doctor based on a double-blind placebo-controlled food challenge – peanut – nuts – sesame – legumes	no	no	yes	no
31	M	XLA	no	no	no	no	no	no	no
32	F	SPAD	no	no	no	yes	no	yes	no
33	M	hypogammaglobulinemia	no	no		yes	no	no	no
34	M	CAPS	yes	yes	no	no	no	no	no
35	M	hyper IgM syndrome	no	no	no	no	no	no	no
36	M	hyper IgM syndrome	no	no	no	not applicable[§]	not applicable[§]	no	no
37	M	CVID	yes	no	no	no	no	no	no
38	M	CVID	yes	yes	diagnosis by doctor based on complaints – lactose	no	no	no	yes
39	M	hypogammaglobulinemia	yes	no	no	no	yes	yes	no
40	F	CVID (Cowden syndrome)	no	no	no	yes	no	no	no
41	M	unknown	yes	no	no	not applicable[§]	not applicable[§]	no	no
42	M	CVID	yes	no	diagnosis by doctor based on complaints – milk	no	no	no	yes
43	F	CVID	yes	no	no	no	no	no	no
44	F	APDS	yes	yes	no	no	yes	yes	yes
45	M	unknown	yes	no	no	no	no	no	no
46	M	hypogammaglobulinemia	yes	no	no	yes		yes	yes
47	M	hypogammaglobulinemia	yes	yes	diagnosis by doctor based on complaints – fruit	yes	yes	yes	yes

Abbreviations: APDS, activated PI3 kinase delta syndrome; CAPS,cryopyrin-associated periodic syndrome;CMC, chronic mucocutaneous candidiasis; CVID, common variable immunodeficiency disorder; F, female; M, male; SPAD, specific polysaccharide antibody deficiency; XLA, X-linked agammaglobulinemia.

[§]As episodic respiratory symptoms, such as wheezing and cough, are very common in children, it is difficult to make the diagnosis of asthma with a high degree of certainty in children aged 1–5 years based on these questions. Therefore, data of patients <5 years were excluded from further analysis.

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Table S2. Current and ever experienced atopic manifestation per adult patient with a primary immunodeficiency disease

Patient number	Sex	Primary immunodeficiency disease	Ever atopic dermatitis	Current atopic dermatitis	AD according to the UK working party diagnostic criteria	Food allergy	sIgE against panel of food allergens (kU/L) + positive allergen(s)	Ever asthma	Current asthma	Bronchodilator reversibility test	Bronchial challenge test with histamine (histamine threshold in mg/mL)	Ever hayfever	Current hayfever	sIgE against panel of inhalation allergands (kU/L) + positive allergen(s)	Smoking status
1	F	IgG1 and IgG3 subclass deficiency	yes	yes				no	yes			yes	yes		non-smoker
2	F	SADNI	no	yes			0.22	yes	yes	no asthma – FEV1/VC: 2.23 SD – increase FEV1: no reversibility	no asthma	yes	yes	0.12	non-smoker
3	M	CVID	no	yes		no		yes	no			no	no		non-smoker
4	F	hypogammaglobulinemia (IgA, IgM, IgG1 and IgG3)	no	no		no		no	no			no	no		unknown
5	F	SADNI	no	yes	no		0.10	yes	yes	no asthma – FEV1/VC: -0.79 SD – increase FEV1: 100 mL and 3.4%	asthma (14)	yes	yes	0.10	current smoker
6	M	IgA and IgG2 subclass deficiency	yes	yes	no		0.10	yes	yes			yes	yes	2.26* – house dust mite (*Dermatophagoides pteronyssinus*): 0.58	non-smoker
7	F	hypogammaglobulinemia (Morbus Steinert)	no	no				yes	no			no	no		non-smoker
8	M	SADNI	no	yes		no		yes	no	no asthma – FEV1/VC: -1.83 SD – increase FEV1: no reversibility		yes	yes		non-smoker
9	M	CVID	no	yes	no	no	0.10	yes	no			no	no	0.10	non-smoker
10	F	hypogammaglobulinemia (IgG and IgM)	no	yes		no		yes	yes			yes	no		ex-smoker
11	M	hyper IgE syndrome	no	no	no			yes	yes	asthma – increase FEV1: 9%	asthma (14.3)	yes	yes		ex-smoker
12	F	Aicardi-Goutieres syndrome	no	no	no	no	0.14	yes	no			no	no	0.31	unknown
13	F	IgG1 and IgG3 subclass deficiency	no				0.10	no	yes			yes	yes	0.10	non-smoker
14	F	CVID	yes	yes	no	no	0.10	yes	no				no	1.27* – cat dander: 3.71	non-smoker
15	F	CVID	no	yes		no	0.10	yes	yes	no asthma – FEV1/VC: 1.15 SD – increase FEV1: no reversibility		no	no	0.10	non-smoker
16	F	CVID	no	no	no	no		no	yes			yes	yes		current smoker

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17	F	CVID	no	no						asthma with airway obstruction – FEV1/VC: 67% – increase FEV1: 10%		yes	yes		ex-smoker
18	F	combined B- en T-cel dysfunction	no	no	no	no	0.11	no	yes	no asthma – FEV1/VC: -0.88 SD – increase FEV1: 130 mL and 3.9%		no	no	0.10	ex-smoker
19	M	CVID	no	yes	no	no	0.10	yes	no	no asthma – FEV1/VC: 1.25 SD – increase FEV1: no reversibility	no asthma	yes	no	12.00* – birch tree pollen: 1.47 – mugwort pollen: 0.62 – grass pollen: 13.60	non-smoker
20	F	IgG3 subclass deficiency	no	yes	no	no		no	yes	no asthma – FEV1/VC: 77% – increase FEV1: no reversibility		no	no	0.34	current smoker
21	M	CVID	no	no		no		no	no			no	no		unknown
22	F	CVID	no	no		no		yes	yes			no	no		non-smoker
23	F	CVID	yes	yes	yes	no		yes	no	asthma with moderately severe airway obstruction – FEV1/VC: -3.78 SD – increase FEV1: 200 mL and 9.5%		yes	yes		ex-smoker
24	F	CMC	no	yes		no		no	no			no	no		non-smoker
25	M	polysaccharide antibody deficiency	no	no		no		no	yes	severe airway obstruction, no asthma – FEV1/VC: -5.47 SD – increase FEV1: no reversibility		no		0.10	ex-smoker
26	F	SADNI	no	yes		no	0.10	no	no			no	no	0.12	current smoker
27	F	CGD	no	no		diagnosis by doctor based on complaints – milk	0.10	no	yes			no	no	0.10	unknown
28	F	DiGeorge syndrome	no	no	no	no	0.10	no	no			no	no	0.10	non-smoker
29	F	CVID	no	yes	no	no	0.10	no	no			no	no	0.10	non-smoker
30	M	IgA, IgG2 and IgG4 subclass deficiency	no	no		no		no	yes	asthma with severe airway obstruction – FEV1/VC: -6.70 SD – increase FEV1: 710 mL and 17.1%		no	no		ex-smoker
31	M	hypogammaglobulinemia (IgG)	no	no		no		yes	yes			yes	yes		ex-smoker
32	F	IgA and IgG4 subclass deficiency	no	yes		no		yes	no			yes	yes	0.10	non-smoker
33	F	SADNI	no	no	yes	no		no	no	no asthma – FEV1/VC: -0.23 SD		no	no	0.38* – cat dander: 0.82	non-smoker

										– increase FEV1: 90 mL and 4.3%					
34	M	CVID	no	yes	no	no		yes	no			no	no		ex-smoker
35	M	IgA subclass deficiency	no	no		no	0.14	yes	no			yes	yes	0.10	non-smoker
36	F	CVID	no	yes		no		yes	no			no	no		non-smoker
37	F	CVID	no	yes		no	0.10	no	no			no	no	0.10	ex-smoker
38	F	CVID (Stiff Person syndrome)	no	no	no	no		yes	no			no	no		non-smoker
39	M	CVID	no	yes		no	0.10	no	yes			yes	yes	1.24* – grass pollen: 0.94	non-smoker
40	F	SADNI	yes	yes	no		0.19	yes	yes		no asthma	no	no	1.21* – house dust mite (*Dermatophagoides pteronyssinus*): 1.27	non-smoker
41	M	CVID	no	no		no	0.10	yes	no			no	no	0.10	unknown
42	M	CVID	no	yes		no	0.10	yes	yes	no asthma – FEV1/VC: -1.01 SD – increase FEV1: 90 mL and 2.9%	asthma (31)	yes	yes	0.10	unknown
43	M	hyper IgE syndrome	yes	yes	yes	diagnosis by doctor based on skin prick test – egg-white – nuts – peanut – fruits – nightshades	64.50* – egg white: 16.40 – milk: 0.56 – peanut: 42.10 – soy: 1.03 – wheat: 1.87	yes	yes			yes	yes	>100* – dog dander: 52.80 – cat dander: 25.80 – rabbit dander: 9.50 – horse dander: 39.70	non-smoker
44	F	CVID	no	yes	no		0.23	yes	yes	no asthma – FEV1/VC: -1.03 SD – increase FEV1: no reversibility	no asthma	yes	yes	0.13	ex-smoker
45	F	SADNI	no	no			0.23	no	no	no asthma – FEV1/VC: -0.81 SD – increase FEV1: 170 mL and 6.5%		yes	yes	0.10	non-smoker
46	F	SADNI	no	yes	no	no		no	yes	airway obstruction, no asthma – FEV1/VC: 68.6% – increase FEV1: 170 mL and 5.6%		yes	yes	0.10	non-smoker
47	F	IgA subclass deficiency	no	no		no	0.10	no	no			yes	yes	0.10	non-smoker
48	F	SADNI	no	no		no	0.10	no	yes	severe airway obstruction, no asthma – FEV1/VC: -4.56 SD – increase FEV1: no reversibility		no	no	0.10	ex-smoker
49	M	CVID	yes	yes	no	no	0.10	no	yes			yes	yes	0.10	non-smoker
50	F	IgG2 subclass deficiency	no	yes	no	no		no	yes			yes	yes		non-

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															smoker
51	M	SADNI	yes	yes	no	diagnosis by doctor based on a double-blind placebo-controlled food challenge – coffee – chocolate		no	yes			yes	yes		non-smoker
52	F	IgG1, IgG2 and IgG4 subclass deficiency	no	yes			0.10	yes	yes			no	no	0.10	current - smoker
53	M	hypogammaglobulinemia (Morbus Waldenström)	no	no		no	0.10	no	no			no	no	0.10	non-smoker
54	F	CVID	no	no	no	no		yes	no			no	no		non-smoker
55	F	hereditary angioedema	no	no		no	0.68* – milk: 1.22	yes	no			no	no	0.10	non-smoker
56		CGD	no	yes	no	no	0.10	no	no			no	no	0.10	current smoker
57	M	SADNI	no	no		no	0.10	yes	no		asthma (24.6)	no	no	1.66* – birch tree pollen: 4.51	non-smoker
58	M	CVID	no	yes	yes			yes	no			yes	no		ex-smoker
59	M	CVID	no	no	no	no		no	no			yes	no		non-smoker
60	M	immune dysregulation syndrome	no	yes		no	0.10	yes	yes	allergic asthma with moderate airway obstruction – FEV1/VC: -2.79 SD – increase FEV1: 390 mL and 21.0%	no asthma	no	no	2.97* – cat dander: 0.52 – house dust mite (*Dermatophagoides pteronyssinus*): 2.86	ex-smoker
61	M	CVID	no	no		no		yes	no			no	no	0.10	non-smoker
62	F	hypogammaglobulinemia	no	no	no	no		no	yes			yes	yes	1.80* – house dust mite (*Dermatophagoides pteronyssinus*): 1.87	non-smoker
63	F	IgG2 subclass deficiency	no	no		no	0.10	yes	yes			no	no	0.10	ex-smoker
64	F	IgG2 subclass deficiency	no	no		no	0.10	no	no			no	no	0.10	non-smoker
65	F	hyper IgE syndrome	no	yes		diagnosis by doctor based on a double-blind placebo-controlled food challenge – milk – egg white – herbs		yes	yes			yes	yes		non-smoker
66	F	CVID	no	no		no		yes	no			yes	yes		unknown
67	F	CVID	no	no				no	no			yes	yes		non-smoker
68	M	SADNI	yes	yes	no	no		no	yes			yes	yes	0.10	non-smoker

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69	M	CVID	no	yes		no	0.10	no	yes	no asthma – FEV1/VC: -0.62 SD – increase FEV1: 230 mL and 4.5%	no asthma	no	no	7.37* unknown allergens	non-smoker
70	M	XLA	no	no	yes	no	0.10	yes	no			no	no	0.10	non-smoker
71	F	CVID	no	no		diagnosis by doctor based on complaints – fish	0.10	yes	yes			no	no	0.10	non-smoker
72	F	hypogammaglobulinemia	no	no	no	no	0.10	yes	no			no	no	0.10	ex-smoker
73	F	IgG2 subclass deficiency	no	yes				yes	yes			yes	no		non-smoker
74	M	IgG2 subclass deficiency	no	yes	no				yes			yes	no		current smoker
75	M	C2 deficiency	no	no		no		yes	no			no	no		ex-smoker
76	M	CVID	no	no		no	0.10	yes	no			no	no	0.10	non-smoker
77	M	CVID	no	yes	no	no	0.10	yes	no			yes	yes	0.10	non-smoker
78	F	CVID	no	yes			0.10	no	yes	no asthma – FEV1/VC: 0.41 SD – increase FEV1: 140 mL and 5.6%		yes	yes	0.10	non-smoker
79	M	CVID	no	no				yes	yes			yes	yes		unknown
80	F	IgG1 subclass deficiency	yes	yes		no		no	no	allergic asthma with severe airway obstruction – FEV1/VC: -4.64 SD – increase FEV1: 210 mL and 13.6%		yes	yes	11.20* – birch tree pollen: 2.55 – grass pollen: 0.61 – dog dander: 0.84 – cat dander: 0.41 – house dust mite (*Dermatophagoides pteronyssinus*): 17.40	non-smoker
81	F	IgG1 subclass deficiency	yes	yes	no	diagnosis by doctor based on complaints – raw vegetables and fruits – pork		yes	yes			yes	yes		non-smoker
82	M	SADNI	no	yes		diagnosis by doctor based on complaints – walnuts		no				no	no		non-smoker
83	F	IgG1 subclass deficiency	no	yes	no	no	0.10	no	yes			yes	yes	0.10	ex-smoker
84	M	Good syndrome	no	no		no	0.10	yes	yes			no	no	0.10	non-smoker
85	M	CVID	yes	yes	yes	no		yes	yes		no asthma	no	no		non-smoker
86	F	hypogammaglobulinemia (Morbus Steinert)	no	no		no		no	no			no	no		non-smoker

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87	F	CVID	no	yes				no	yes			yes	yes		current smoker
88	F	hypogammaglobulinemia	no	yes	no	no		yes	no			no	no		ex-smoker
89	M	CVID	no		no	no	0.10	no	yes			yes	yes	0.10	ex-smoker
90	M	CVID	no	no	no	no	0.10	yes	no	no asthma – FEV1/VC: -1.35 SD – increase FEV1: no reversibility	no asthma	yes	yes	0.10	non-smoker
91	xM	hypogammaglobulinemia	yes	yes		no		no	no			no			ex-smoker
92	xF	CVID	yes	yes		no		yes	yes			yes	yes		non-smoker
93	F	CVID	no	no	no	no	0.10	yes	yes	moderate airway obstruction, no asthma – FEV1/VC: -2.32 SD – increase FEV1: 100 mL and 4.3%		no	no	0.10	non-smoker
94	F	CMC	no	no		no		no	no			no	no		ex-smoker
95	M	IgA, IgG2 and IgG4 subclass deficiency	no	no		no	0.10	yes	no	no asthma – FEV1/VC: -1.01 SD – increase FEV1: 11%		yes	yes	0.10	non-smoker
96	F	CVID	yes	yes	no	diagnosis by doctor based on complaints – fungi – yeasts		no	no			no	no		non-smoker
97	M	combined B- and T- cel dysfunction	no	no		no		no	no			yes	yes		non-smoker
98	M	hypogammaglobulinemia	yes	yes	no	diagnosis by doctor based on a double-blind placebo-controlled food challenge – milk		no	no			no	no		non-smoker
99	M	CVID	no	no		no		yes	no	no asthma – FEV1/VC: -0.23 SD – increase FEV1: 50 mL and 1.1%		no	no	0.10	ex-smoker
100	F	hypogammaglobulinemia (IgG1 and IgG4)	no	no				yes	yes			no	no		current smoker
101	F	CVID	no	no	no	no	0.16	no	yes	no asthma – FEV1/VC: -0.88 SD – increase FEV1: 190 mL and 5.9%	no asthma	yes	yes	0.46* – house dust mite (*Dermatophagoides pteronyssinus*): 0.47	non-smoker
102	F	SADNI	no	yes		no	0.10	yes	no			no	no	0.10	ex-smoker
103	F	hypogammaglobulinemia	no	yes	no	no	0.10	yes	yes			yes	yes	0.10	ex-smoker
104	M	XLA	no	no		no		yes	no			no	no		unknown
105	M	XLA	no	no	no	no	0.10	no	no	moderately severe airway obstruction, no asthma		no	no	0.10	non-smoker

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										– FEV1/VC: -3.00 SD – increase FEV1: 130 mL and 4.2%					
106	M	CVID	no	no	no	no		yes	no			no	no		non-smoker
107	M	CVID	no	no	no	no		yes	yes	no asthma – FEV1/VC: -0.67 SD – increase FEV1: 200 mL and 5.9%		yes	yes	0.10	non-smoker
108	F	CVID		no	no	no	0.10	yes	yes	no asthma – FEV1/VC: -0.51 SD – increase FEV1: 50 mL and 1.2%		yes	yes	0.10	non-smoker
109	F	cellular immunodeficiency	no	yes		no		yes	no	moderately severe airway obstruction, no asthma – FEV1/VC: -3.20 SD – increase FEV1: 150 mL and 5.4%		no	no	15.00* – grass pollen: 16.90 – house dust mite (*Dermatophagoides pteronyssinus*): 4.08	ex-smoker
110	F	CVID	no	no	no	no		yes	no	asthma with airway obstruction – FEV1/VC: 41.3% – increase FEV1: 190 mL and 19.4%		yes	yes		non-smoker
111	M	hypogammaglobulinemia (IgG, IgG1 and IgG2)	no	yes		no		yes	yes			yes	no		ex-smoker
112	M	XLA	no	no	no	no		no	yes			yes	yes	0.10	non-smoker
113	F	CVID	no	no		no	0.10	yes	no	no asthma – FEV1/VC: -0.87 SD – increase FEV1: 10 mL and 0.6%		yes	yes	0.10	ex-smoker
114	F	hypogammaglobulinemia (IgG, IgG1 and IgG2)	no	yes		no	0.10	no	yes			yes	yes	0.32	non-smoker
115	F	IgG1 and IgG2 subclass deficiency	no	no	no	no	0.10	yes	yes			yes	yes	0.10	current smoker
116	M	IgG2 subclass deficiency	yes	yes	no	no	0.10	yes	yes			yes	yes	0.10	non-smoker
117	F	hypogammaglobulinemia (IgG1, IgG2 and IgG3)	no	yes	no		0.10	yes	yes	moderate airway obstruction, no asthma – FEV1/VC: -2.90 SD – increase FEV1: 100 mL and 3.6%		yes	yes	0.10	ex-smoker
118	xM	Good syndrome	no	no		no		no	yes			no	no		ex-smoker
119	M	hyper IgE syndrome	no	yes	no	no	1.97* – egg white: 1.77 – milk: 0.76 – peanut: 0.48 – wheat: 0.31	yes	yes	moderately severe airway obstruction, no asthma – FEV1/VC: -3.70 SD – increase FEV1: 140 mL and 9.7%		no	no	11.90* – mugwort pollen: 0.40 – grass pollen: 2.61 – dog dander: 0.85 – cat dander: 5.94 – house dust mite (*Dermatophagoides pteronyssinus*):13.20	non-smoker

120	F	CVID	no	yes		no		no	no			no	no	0.10	ex-smoker
121	F	CVID	yes	yes		no		yes	yes			yes	yes		unknown
122	M	CVID	no	no	no	no	0.10	yes	no			no	no	0.10	non-smoker
123	M	SADNI	no	no		no		yes	yes	airway obstruction, no asthma – FEV1/VC: 69% – increase FEV1: no reversibility		yes	yes	0.10	non-smoker
124	xF	IgG2 subclass deficiency	no	yes		no		yes	yes			no	no		current smoker
125	F	IgG2 subclass deficiency	no	yes	no	diagnosis by doctor based on complaints – nuts	0.96* – peanut: 2.22 – soy: 0.56 – wheat: 0.83 – nut mix: 3.95 – nut mixture: 0.92 – pistachio nut: 0.87 – almond: 1.61	no	no			yes	yes	6.32* – birch tree pollen: 6.60 – mugwort pollen: 0.45 – grass pollen: 5.98 – cat dander: 0.31	non-smoker
126	M	XLA	yes	yes	yes		0.10	yes	no	moderate airway obstruction, no asthma – FEV1/VC: -2.29 SD – increase FEV1: 180 mL and 5.1%		no	no	0.10	non-smoker
127	F	IgG1 subclass deficiency	no	yes		no	0.10	no	yes			yes	yes	0.10	ex-smoker
128	F	IgG1 en IgG2 subclass deficiency	no	no		no		no	no			yes	yes		current smoker
129	F	IgG1 en IgG3 subclass deficiency	no	yes	no	no	0.10	no	no			yes	yes	0.10	non-smoker
130	F	hypogammaglobulinemia (IgG, IgG1, IgG2, IgA and IgM)	no	yes	no	no	0.10	no	yes	no asthma – FEV1/VC: -0.43 SD – increase FEV1: 40 mL and 1.3%	no asthma	no	no	0.10	non-smoker
131	F	IgG1 and IgG3 subclass deficiency	no	yes		diagnosis by doctor based on complaints – fish – shellfish – apple	0.10		yes			yes	yes	3.35* – birch tree pollen: 1.19 – house dust mite (*Dermatophagoides pteronyssinus*): 2.80	non-smoker
132	F	IgG1 and IgG4 subclass deficiency	no	yes		no	0.10	yes	yes	severe airway obstruction, no asthma – FEV1/VC: -3.45 SD – increase FEV1: 140 mL and 6.2%		yes	yes	0.10	current smoker
133	F	SADNI	no	no	no	no	0.25	yes	no	no asthma – FEV1/VC: -1.18 SD – increase FEV1: 30 mL and 1.3%	no asthma	no	no	0.10	non-smoker
134	F	IgG1 and IgG3 subclass deficiency	no	no		no	0.10	no	no	no asthma – FEV1/VC: 77% – increase FEV1: no		yes	yes	1.09* – birch tree pollen: 3.15	non-smoker

										reversibility					
135	F	IgA deficiency	no			no		yes	no			yes	yes		current smoker
136	F	TRAPS	no	no		no		no	no			yes	yes		current smoker
137	F	TRAPS	no	no		no		yes	no			no	no		non-smoker
138	F	agammaglobulinemie	no	no	no	no	0.10	no	no	no asthma – FEV1/VC: -1.52 SD – increase FEV1: no reversibility		yes	no	0.10	non-smoker
139	F	hypogammaglobulinemia (IgG2)	no	yes	yes	no		no	no			no	no		non-smoker
140	F	CVID	no	no	no	no		no	yes	no asthma – FEV1/VC: -0.80 SD – increase FEV1: 150 mL and 4.1%		no	no		non-smoker
141	F	hypogammaglobulinemia (Morbus Steinert)	no	yes	no	no	0.16	yes	no			yes	no	0.10	non-smoker
142	M	hyper IgM syndrome	no	no		no		no	no			no	no		non-smoker
143	F	hyper IgE syndrome	no	yes	yes	no	56.90* – milk: 66.20	no	yes	moderate airway obstruction, no asthma – FEV1/VC: -2.76 SD – increase FEV1: 180 mL and 11.5%		yes	yes	1.38* – birch tree pollen: 0.64 – mugwort pollen: 0.70 – grass pollen: 0.84 – dog dander: 3.79 – house dust mite (*Dermatophagoides pteronyssinus*): 0.66	non-smoker
144	M	FMF	no	no	no	no	0.10	yes	no			no	no	0.10	non-smoker
145	F	IgG2 and IgG4 subclass deficiency	no	no		no		yes	no			no	no		non-smoker
146	F	SADNI	no	no	no	no		no	no			no	no	0.10	non-smoker
147	M	CVID	no	no	no			yes	no			no	no		non-smoker
148	F	IgG2 subclass deficiency	no	no		diagnosis by doctor based on complaints – milk		yes	no			no	no		current smoker
149	M	SADNI	yes	yes	no	no		yes	yes	mild airway obstruction, no asthma – FEV1/VC: -1.81 SD – increase FEV1: no reversibility	no asthma	no	no		non-smoker
150	M	CVID	no	no	no	no	0.10	no	no	no asthma – FEV1/VC: 0.01 SD – increase FEV1: 150 mL and 3.8%	no asthma	yes	no	1.81* – grass pollen: 1.13 – house dust mite (*Dermatophagoides pteronyssinus*): 1.29	non-smoker
151	F	CVID	no	no		no	0.10	yes	no			no	no	0.10	non-smoker
152	F	CVID	no	no		no	0.10	no	no			no	no	0.10	non-

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															smoker
153	M	IgG3 subclass deficiency	no	yes	no	no	0.10	yes	yes			no	no	0.15	ex-smoker
154	F	SADNI	no	no		no		yes	no			no	no	0.10	ex-smoker
155	M	SADNI	no	no		no	0.10	no	yes		asthma (7.5)	yes	yes	0.10	non-smoker
156	F	CVID	no	yes		diagnosis by doctor based on a double-blind placebo-controlled food challenge – wheat – soy – milk	0.30	yes	yes		no asthma	yes	yes	5.62* – grass pollen: 0.59 – dog dander: 0.48 – house dust mite (*Dermatophagoides pteronyssinus*): 4.99	non-smoker
157	M	XLA	no	no	no	no	0.10	no	yes	non-allergic asthma with mild airway obstruction – FEV1/VC: -1.87 SD – increase FEV1: 360 mL and 9.4%		no	no	0.10	unknown
158	F	SADNI	no	no		no		yes	no			no	no		non-smoker
159	M	SADNI	no	yes	no	no	0.10	no	yes			no	no	1.08* – house dust mite (*Dermatophagoides pteronyssinus*): 1.37	ex-smoker
160	xM	CVID (Jacobsen syndrome)	yes	yes				yes	no			yes	yes		non-smoker
161	F	CMC	no	no	no		0.10	yes	yes	no asthma – FEV1/VC: 0.08 SD – increase FEV1: no reversibility		yes	yes	0.10	non-smoker
162	F	IgA deficiency	no	yes				yes	yes			no	no	0.10	ex-smoker
163	F	hypogammaglobulinemia (Good syndrome)	no	yes	no		0.10	no	no			no	no	0.10	unknown
164	M	CVID	no	yes	no	no		yes	yes	no asthma – FEV1/VC: 0.37 SD – increase FEV1: 120 mL and 2.6%	no asthma	yes	yes	0.10	non-smoker
165	M	IgG2 subclass deficiency	no	no		no	0.10	no	no	allergic asthma with moderate airway obstruction – FEV1/VC: -2.41 SD – increase FEV1: 470 mL and 13.6%		yes	no	20.20* – birch tree pollen: 28.10 – grass pollen: 1.52 – dog dander: 0.40 – cat dander: 1.17 – house dust mite (*Dermatophagoides pteronyssinus*): 16.00	ex-smoker
166	M	IgG1 and IgG3 subclass deficiency	no	no		no		no	yes			yes	yes		ex-smoker
167	M	IgG2 subclass deficiency		yes		no		yes	no	no asthma – FEV1/VC: -0.63 SD	no asthma	no	no		non-smoker

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										– increase FEV1: 340 mL and 8.3%					
168	F	hypogammaglobulinemia (IgG)	no	yes		diagnosis by doctor based on a double-blind placebo-controlled food challenge – soy – nuts – peanut – spinach	0.12	no	yes	no asthma – FEV1/VC: -0.51 SD – increase FEV1: 140 mL and 4.3%		yes	yes	14.10* – birch tree pollen: 32.60 – mugwort pollen: 0.37 – grass pollen: 8.79 – dog dander: 0.54	non-smoker
169	F	CVID	no	yes	no	no	0.10	no	yes	no asthma – FEV1/VC: -1.40 SD – increase FEV1: no reversibility		yes	yes	0.10	non-smoker
170	xM	hypogammaglobulinemia (IgG)	no	yes				no	yes			yes	yes		current smoker
171	F	hypogammaglobulinemia (Morbus Steinert)	no	no		no		yes	no			no	no		ex-smoker
172	F	IgG2, IgG3 and IgG3 subclass deficiency	no	yes	no	no	0.10	yes	yes			yes	yes	0.10	current smoker
173	M	SADNI	no	no		no		no	no			no	no		non-smoker
174	M	SADNI	no	no				yes	no	no asthma – FEV1/VC: -1.54 SD – increase FEV1: 60 mL and 2.0%		yes	yes	0.10	non-smoker
175	F	CVID	no	no	no	no	0.10	yes	no			no	no	0.10	ex-smoker
176	F	CVID	no	yes	no	no		no	no		no asthma	yes	yes		current smoker
177	M	hypogammaglobulinemia (Good syndrome)	no	no	no	no	0.10	no	no			no	no	0.10	ex-smoker
178	F	hypogammaglobulinemia (IgG1)	no	no	no	no	0.10	no	no			yes	yes	0.10	current smoker
179	F	SADNI	no	yes		diagnosis by doctor based on complaints – mint	0.11	yes	yes	no asthma – FEV1/VC: -0.74 SD – increase FEV1: 50 mL and 2.2%		no	no	0.10	current smoker
180	F	IgG1 and IgG4 subclass deficiency	no				0.10	yes	no			yes	yes	0.10	ex-smoker
181	F	TRAPS	no	no		no		no	no			no	no		non-smoker
182	F	CVID	no	no	no	no	0.10	yes	yes			yes	yes	0.10	non-smoker
183	F	hyper IgM syndrome	no	no		no		yes	no			yes	yes	0.10	non-smoker
184	M	CMC	no	yes	no	no	0.10	yes	no			no	no	0.10	non-smoker
185	F	CMC	no	no	no		0.10	yes	no			no	no	0.10	ex-smoker
186	M	CVID	no	yes		no		no	no			no	no		non-smoker
187	F	CVID	no	yes		no	0.10	no	yes	no asthma		yes	yes	0.10	unknown

										– FEV1/VC: 0.37 SD – increase FEV1: no reversibility					
188	M	CVID	no	no		no	0.10	yes	yes			no	no	0.10	ex-smoker
189	F	CVID	no	yes	yes	diagnosis by doctor based on complaints – food additives – E-numbers		yes	no			yes	yes		current smoker
190	M	hyper IgM syndrome	no	yes		no		yes	yes			no	no		ex-smoker
191	F	CVID	no	yes	no		0.10	yes	yes			no	no	0.45* – grass pollen: 0.46	non-smoker
192	F	hypogammaglobulinemia (IgG, Ig1 en IgG3)	no	yes	yes	no		yes	yes	no asthma – FEV1/VC: -1.36 SD – increase FEV1: no reversibility	asthma (16.5)	yes	yes	0.10	ex-smoker
193	F	hypogammaglobulinemia (B-cel)	no	yes	no	no		no	yes	no asthma – FEV1/VC: 0.79 SD – increase FEV1: 110 mL and 3.7%	no asthma	yes	yes	0.10	non-smoker
194	M	CVID	no	no	no	no	0.10	yes	no			no	no	0.10	non-smoker
195	F	SADNI	no	yes	no	no	0.10	no	no	no asthma – FEV1/VC: -1.35 SD – increase FEV1: 70 mL and 3.0%		no	no	0.10	non-smoker
196	M	CVID	no	no	no	diagnosis by doctor based on a double-blind placebo-controlled food challenge – cow's milk – egg white	0.10	no	no			yes	yes	0.10	current smoker
197	M	CVID	no	no		no	0.10	no	no			no	no	0.10	current smoker
198	F	CVID	no	no		no		no	yes			no	no		unknown
199	M	CVID	yes	yes	no	no	0.10	no	no			yes	yes	0.10	ex-smoker
200	M	IgG1 subclass deficiency	no	yes		diagnosis by doctor based on complaints – pork		yes	yes			yes	yes		current smoker
201	F	CVID	no	yes	no	no	0.10	yes	no			yes	yes	0.10	non-smoker
202	M	IgA deficiency	no	no		no		no	yes	no asthma – FEV1/VC: -1.33 SD – increase FEV1: 210 mL and 6.1%		yes	yes	0.10	ex-smoker
203	F	CVID	no	no	no	no		yes	no			yes	yes		non-smoker
204	M	hypogammaglobulinemia	no	yes	no	no	0.10	yes	no			yes	yes	0.10	non-

		(IgM, IgA, IgG; ICF 2 syndrome)													smoker
205	F	CVID	no	no	no	no		yes	no	non-allergic asthma with moderate airway obstruction – FEV1/VC: -2.36 SD – increase FEV1: 270 mL and 10.7%	no asthma	yes	yes	0.10	non-smoker
206	F	IgG4 subclass deficiency	no	no	no			yes	no			no	no		non-smoker

Abbreviations: AD, atopic dermatitis; APDS, activated PI3 kinase delta syndrome; CAPS, cryopyrin-associated periodic syndrome; CMC, chronic mucocutaneous candidiasis; CVID, common variable immunodeficiency disorder; F, female; FMF, familial Mediterranean fever; M, male; SADNI, selective antibody deficiency with normal immunoglobulins; SPAD, specific polysaccharide antibody deficiency; TRAPS, tumor necrosis factor receptor-associated periodic syndrome; XLA, X-linked agammaglobulinemia.

Table S3. Current and ever experienced atopic manifestation per adult partner-control

Patient number	Sex	Ever atopic dermatitis	Current atopic dermatitis	Food allergy	Ever asthma	Current asthma	Ever hayfever	Current hayfever
1	M	no	no	no	no	no	no	no
2	M	no	yes	no	no	no	yes	yes
3	F	no	no	no	no	no	no	no
4	M	no	no	no	no	no	no	no
5	M	no	no	no	no	no	yes	no
6	M	no	no	no	no	no	no	no
7	M	yes	no	no	no	no	no	no
8	F	no	no	no	no	no	no	no
9	M	no	no	no	no	no	no	no
10	M	no	no	no	no	no	no	no
11	M	no	no	no	no	no	no	no
12	F	no	no	no	no	no	no	no
13	F	no	no	no	no	yes	no	no
14	M	no	no	no	no	no	no	no
15	F	no	no	no	no	no	yes	yes
16	F	no	no	no	no	no	no	no
17	F	yes	no		no	no	no	no
18	F	yes	no	no	yes	yes	no	no
19	M	no	no	no	no	no	no	no
20	M	no	no	no	no	no	no	no
21	F	no	no	no	no	no	yes	no
22	F	no	no	no	yes	yes	no	yes
23	F	yes	no	no	yes	no	no	no
24	M	no	no	no	no	no	no	no
25	M	no	yes	no	yes	no	no	no
26	F	yes	no	no	no	no	no	no
27	M	yes		no	no	no	no	no
28	F	no	no	no	no	no	yes	no
29	M	no	no	no	yes	yes	yes	yes
30	M	yes	yes	no	no	no	no	no
31	M	no	no	no	no	no	no	no
32	M		no					
33	M	yes	no		no	no	no	yes
34	M	no	no	no	no	no	no	no
35	M	no	no	no	no	no	no	no
36	F	no	no	no	no	no	yes	no
37	F	no	no	no	no	no	no	yes
38	F	no	no	no	no	no	no	no
39	F	no	no	no	no	no	yes	no
40	M	no	no	no			no	no
41	F	yes	no	diagnosis by doctor based on complaints	no	no	yes	yes
42	F	yes	no	no	no	no	no	yes
43	M	no	no	no	no	no	yes	yes
44	M	no	no	no	no	no	no	no

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45	M	no	no	no	no	no	yes	no
46	M	yes	no	no	no	yes	no	no
47	F	no	no	no	no	no	no	no
48	M	yes	no	no	no	no	no	no
49	M	yes	no	no	no	no	no	no
50	F	yes	no	no	yes	yes	no	no
51	M	no	no	no	no	no	no	no
52	M	yes	no	no	yes	no	yes	no
53	F	no	yes	no	no	no	no	no
54	M	no	no	no	no	no	no	no
55	M	no	no	no	yes	yes	no	no
56	M	no	no	no	no	no	no	no
Abbreviations: F, female; M, male.								

Appendix 1

	Current	**Ever experienced**
Atopic dermatitis*	Positive answers to both the questions: - "Have you (has your child) had this itchy rash at any time in the past 12 months?" - "Has this itchy rash at any time affected any of the following places: the folds of the elbows; behind the knos; in front of the ankles; under the buttocks; or around the neck, ears, or eyes?"	Positive response to the question: - "Have you (has your child) ever had eczema?"
Asthma*	Positive answer to the question: - "Have you (has your child) had wheezing or whistling in the chest in the past 12 months?"	Positive response to the question: - "Have you (has your child) ever had asthma?"
Allergic rhinitis*	Positive answers to both the questions: - "In the past 12 months, have you (has your child) had a problem with snozing or a runny or blocked nose when you (he/she) did not have a cold or the flu?" - "In the past 12 months, has this nose problem been accompanied by itchy watery eyes?"	Positive response to the question: - "Have you (has your child) ever had hayfever?"
*According to The Phase Three Core Questionnaire of the International Study of Asthma and Allergies in Childhood (1).		

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Appendix 2

Questionnaire pediatric patient 0-11 years

Questionnaire skin disorders in children with a primary immunodeficiency disease

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Study number: ……

Date: ….. / …… / ……

Please complete all questions by the parent(s) and/or caregiver(s)

Primary immunodeficiency disease

1. **What was the your child's age when he/she developed symptoms that fit the diagnosis of primary immunodeficiency disease for the first time? Could you please describe these symptoms?**

   ….. year

   Symptom(s): ….

2. **What was the your child's age when the primary immunodeficiency disease was diagnosed?**

   ….. year

Asthma

1.	**Has your child ever had wheezing or whistling in the chest at any time in the past?**	□ yes □ no*(continue to question 6)*
2.	**Has your child has wheezing or whistling in the chest in the past 12 months?**	□ yes □ no *(continue to question 6)*
3.	**How many attacks of wheezing has your child had in the past 12 months?**	□ none □ 1 to 3 □ 4 to 12 □ more than 12
4.	**In the past 12 months, how often, on average, has your child's sleep been disturbed due to wheezing?**	□ never woken with wheezing □ less than one night per week □ one or more nights per week
5.	**In the past 12 months, has wheezing ever been severe enough to limit your child's speech to only one or two words at a time between breaths?**	□ yes □ no
6.	**Has your child ever had asthma?**	□ yes □ no
7.	**In the past 12 months, has your child's chest sounded wheezy during or after exercise?**	□ yes □ no
8.	**In the past 12 months, has your child had a dry cough at night, apart from a cough associated with a cold or chest infection?**	□ yes □ no

Rhinitis

1. Has your child ever had a problem with sneezing, or a runny, or blocked nose when he/she did not have a cold or the flu?

□ yes
□ no *(continue to question 6)*

2. In the past 12 months, has your child had a problem with sneezing, or a runny, blocked nose when he/she did not have a cold or the flu?

□ yes
□ no *(continue to question 6)*

3. In the past 12 months, has this nose problem been accompanied by itchy-watery eyes?

□ yes
□ no

4. In which of the past 12 months did this nose problem occur? *Please tick any which apply*

□ January
□ February
□ March
□ April
□ May
□ June
□ July
□ Augustus
□ September
□ October
□ November
□ December

5. In the past 12 months, how much did this nose problem interfere with your child's daily activities?

□ not at all
□ a little
□ a moderate amount
□ a lot

6. Has your child ever had hayfever?

□ yes
□ no

Eczema

1.	**Has your child ever had an itchy rash which was coming and going for at least six months?**	□ yes □ no *(continue to question 7)*
2.	**Has your child had this itchy rash at any time in the past 12 months?**	□ yes □ no *(continue to question 7)*
3.	**Has this itchy rash at any time affected any of the following places: the folds of the elbows, behind the knees, in front of the ankles, under the buttocks, or around the neck, ears or eyes?**	□ yes □ no
4.	**At what age did this itchy rash first occur?**	□ under 2 years □ age 2-4 years □ age 5 or more
5.	**Has this rash cleared completely at any time during the past 12 months?**	□ yes □ no
6.	**In the past 12 months, how often, on average, has your child been kept awake at night by this itchy rash?**	□ never in the past 12 months □ less than one night per week □ one or more nights per week
7.	**Has your child ever had eczema?**	□ yes □ no

Additional atopy questions

1. **From what age does your child have symptoms suitable for asthma?**

   ….. year

   □ not applicable

2. **From what age does your child have symptoms suitable for hayfever?**

   ….. year

   □ not applicable

3. **From what age does your child have symptoms suitable for eczema?**

   ….. year

   □ not applicable

4. **Does your child have a food allergy?**
   - □ yes, specify…
   - □ no
   - □ unknown

5. **What was your child's age at diagnosis of the food allergy?**

   ….. year

6. **Who diagnosed the food allergy?**
   - □ parent(s) and/or caregiver(s) based on complaints
   - □ a doctor based on complaints
   - □ a doctor based on a double-blind placebo-controlled food challenge
   - □ other, specify…
   - □ unknown

Appendix 3

Questionnaire pediatric patient 12-17 years

Questionnaire skin disorders in children with a primary immunodeficiency disease

---

Study number: ……

Date: ….. / …… / ……

Please indicate per question who has answered the question; patient (child), parent(s) and/or caregiver(s) or together

Primary immunodeficiency disease

1. **What was the your child's age when he/she developed symptoms that fit the diagnosis of primary immunodeficiency disease for the first time? Could you please describe these symptoms?**

   ….. year

   Symptom(s): ….

2. **What was the your child's age when the primary immunodeficiency disease was diagnosed?**

   ….. year

Asthma

□ patient (child)
□ parent(s) and/or caregiver(s)
□ together

1.	**Have you ever had wheezing or whistling in the chest at any time in the past?**	□ yes □ no *(continue to question 6)*
2.	**Have youhad wheezing or whistling in the chest in the past 12 months?**	□ yes □ no *(continue to question 6)*
3.	**How many attacks of wheezing have you had in the past 12 months?**	□ none □ 1 to 3 □ 4 to 12 □ more than 12
4.	**In the past 12 months, how often, on average, have your sleep been disturbed due to wheezing?**	□ never woken with wheezing □ less than one night per week □ one or more nights per week
5.	**In the past 12 months, has wheezing ever been severe enough to limit your speech to only one or two words at a time between breaths?**	□ yes □ no
6.	**Have you ever had asthma?**	□ yes □ no
7.	**In the past 12 months, have your chest sounded wheezy during or after exercise?**	□ yes □ no
8.	**In the past 12 months, have you had a dry cough at night, apart from a cough associated with a cold or chest infection?**	□ yes □ no

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Rhinitis

□ patient (child)
□ parent(s) and/or caregiver(s)
□ together

1. Have youever had a problem with sneezing, or a runny, or blocked nose when you did not have a cold or the flu?
□ yes
□ no *(continue to question 6)*

2. In the past 12 months, have youhad a problem with sneezing, or a runny, blocked nose when you did not have a cold or the flu?
□ yes
□ no *(continue to question 6)*

3. In the past 12 months, has this nose problem been accompanied by itchy-watery eyes?
□ yes
□ no

4. In which of the past 12 months did this nose problem occur? *Please tick any which apply*
□ January
□ February
□ March
□ April
□ May
□ June
□ July
□ Augustus
□ September
□ October
□ November
□ December

5. In the past 12 months, how much did this nose problem interfere with your daily activities?
□ not at all
□ a little
□ a moderate amount
□ a lot

6. Have youever had hayfever?
□ yes
□ no

Eczema

□ patient (child)
□ parent(s) and/or caregiver(s)
□ together

1. Have youever had an itchy rash which was coming and going for at least six months?
□ yes
□ no *(continue to question 6)*

2. Have youhad this itchy rash at any time in the past 12 months?
□ yes
□ no *(continue to question 6)*

3. Has this itchy rash at any time affected any of the following places: the folds of the elbows, behind the knees, in front of the ankles, under the buttocks, or around the neck, ears or eyes?
□ yes
□ no

4. Has this rash cleared completely at any time during the past 12 months?
□ yes
□ no

5. In the past 12 months, how often, on average, have youbeen kept awake at night by this itchy rash?
□ never in the past 12 months
□ less than one night per week
□ one or more nights per week

6. Have youever had eczema?
□ yes
□ no

Additional atopy questions

- □ patient (child)
- □ parent(s) and/or caregiver(s)
- □ together

1. From what age do you have symptoms suitable for asthma?

….. year

- □ not applicable

2. From what age do youhave symptoms suitable for hayfever?

….. year

- □ not applicable

3. From what age do youhave symptoms suitable for eczema?

….. year

- □ not applicable

4. Do you have a food allergy?

- □ yes, specify…
- □ no
- □ unknown

5. What was your age at diagnosis of the food allergy?

….. year

6. Who diagnosed the food allergy?

- □ parent(s) and/or caregiver(s) based on complaints
- □ a doctor based on complaints
- □ a doctor based on a double-blind placebo-controlled food challenge
- □ other, specify…
- □ unknown

Appendix 4

Questionnaire adult patients

Questionnaire skin disorders in adults with a primary immunodeficiency disease

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Study number: ……

Date: ….. / …… / ……

Primary immunodeficiency disease

1. **What was your age when you developed symptoms that fit the diagnosis of primary immunodeficiency disease for the first time? Could you please describe these symptoms?**

   ….. year

   Symptom(s): ….

2. **What was your age when the primary immunodeficiency disease was diagnosed?**

   ….. year

Asthma

1.	**Have you ever had wheezing or whistling in the chest at any time in the past?**	□ yes □ no *(continue to question 6)*
2.	**Have youhad wheezing or whistling in the chest in the past 12 months?**	□ yes □ no *(continue to question 6)*
3.	**How many attacks of wheezing have you had in the past 12 months?**	□ none □ 1 to 3 □ 4 to 12 □ more than 12
4.	**In the past 12 months, how often, on average, have your sleep been disturbed due to wheezing?**	□ never woken with wheezing □ less than one night per week □ one or more nights per week
5.	**In the past 12 months, has wheezing ever been severe enough to limit your speech to only one or two words at a time between breaths?**	□ yes □ no
6.	**Have you ever had asthma?**	□ yes □ no
7.	**In the past 12 months, have your chest sounded wheezy during or after exercise?**	□ yes □ no
8.	**In the past 12 months, have you had a dry cough at night, apart from a cough associated with a cold or chest infection?**	□ yes □ no

Rhinitis

1. **Have you ever had a problem with sneezing, or a runny, or blocked nose when you did not have a cold or the flu?**
- □ yes
- □ no *(continue to question 6)*

2. **In the past 12 months, have you had a problem with sneezing, or a runny, blocked nose when you did not have a cold or the flu?**
- □ yes
- □ no *(continue to question 6)*

3. **In the past 12 months, has this nose problem been accompanied by itchy-watery eyes?**
- □ yes
- □ no

4. **In which of the past 12 months did this nose problem occur?** *Please tick any which apply*
- □ January
- □ February
- □ March
- □ April
- □ May
- □ June
- □ July
- □ Augustus
- □ September
- □ October
- □ November
- □ December

5. **In the past 12 months, how much did this nose problem interfere with your daily activities?**
- □ not at all
- □ a little
- □ a moderate amount
- □ a lot

6. **Have you ever had hayfever?**
- □ yes
- □ no

Eczema

1.	**Have you ever had an itchy rash which was coming and going for at least six months?**	□ yes □ no *(continue to question 6)*
2.	**Have you had this itchy rash at any time in the past 12 months?**	□ yes □ no *(continue to question 6)*
3.	**Has this itchy rash at any time affected any of the following places: the folds of the elbows, behind the knees, in front of the ankles, under the buttocks, or around the neck, ears or eyes?**	□ yes □ no
4.	**Has this rash cleared completely at any time during the past 12 months?**	□ yes □ no
5.	**In the past 12 months, how often, on average, have you been kept awake at night by this itchy rash?**	□ never in the past 12 months □ less than one night per week □ one or more nights per week
6.	**Have you ever had eczema?**	□ yes □ no

Additional atopy questions

1. **From what age do you have symptoms suitable for asthma?**

   ….. year

   □ not applicable

2. **From what age do you have symptoms suitable for hayfever?**

   ….. year

   □ not applicable

3. **From what age do you have symptoms suitable for eczema?**

   ….. year

   □ not applicable

4. **Do you have a food allergy?**
   - □ yes, specify…
   - □ no
   - □ unknown

5. **What was your age at diagnosis of the food allergy?**

   ….. year

6. **Who diagnosed the food allergy?**
   - □ parent(s) and/or caregiver(s) based on complaints
   - □ a doctor based on complaints
   - □ a doctor based on a double-blind placebo-controlled food challenge
   - □ other, specify…
   - □ unknown

Appendix 5

Questionnaire adult partner-controls

Questionnaire skin disorders in partners of adults with a primary immunodeficiency disease

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Study number: ……

Date: ….. / …… / ……

Asthma

1.	**Have you ever had wheezing or whistling in the chest at any time in the past?**	□ yes □ no *(continue to question 6)*
2.	**Have youhad wheezing or whistling in the chest in the past 12 months?**	□ yes □ no *(continue to question 6)*
3.	**How many attacks of wheezing have you had in the past 12 months?**	□ none □ 1 to 3 □ 4 to 12 □ more than 12
4.	**In the past 12 months, how often, on average, have your sleep been disturbed due to wheezing?**	□ never woken with wheezing □ less than one night per week □ one or more nights per week
5.	**In the past 12 months, has wheezing ever been severe enough to limit your speech to only one or two words at a time between breaths?**	□ yes □ no
6.	**Have you ever had asthma?**	□ yes □ no
7.	**In the past 12 months, have your chest sounded wheezy during or after exercise?**	□ yes □ no
8.	**In the past 12 months, have you had a dry cough at night, apart from a cough associated with a cold or chest infection?**	□ yes □ no

Rhinitis

1. **Have you ever had a problem with sneezing, or a runny, or blocked nose when you did not have a cold or the flu?**
- □ yes
- □ no *(continue to question 6)*

2. **In the past 12 months, have you had a problem with sneezing, or a runny, blocked nose when you did not have a cold or the flu?**
- □ yes
- □ no *(continue to question 6)*

3. **In the past 12 months, has this nose problem been accompanied by itchy-watery eyes?**
- □ yes
- □ no

4. **In which of the past 12 months did this nose problem occur?** *Please tick any which apply*
- □ January
- □ February
- □ March
- □ April
- □ May
- □ June
- □ July
- □ Augustus
- □ September
- □ October
- □ November
- □ December

5. **In the past 12 months, how much did this nose problem interfere with your daily activities?**
- □ not at all
- □ a little
- □ a moderate amount
- □ a lot

6. **Have you ever had hayfever?**
- □ yes
- □ no

Eczema

1.	**Have you ever had an itchy rash which was coming and going for at least six months?**	□ yes □ no *(continue to question 6)*
2.	**Have you had this itchy rash at any time in the past 12 months?**	□ yes □ no *(continue to question 6)*
3.	**Has this itchy rash at any time affected any of the following places: the folds of the elbows, behind the knees, in front of the ankles, under the buttocks, or around the neck, ears or eyes?**	□ yes □ no
4.	**Has this rash cleared completely at any time during the past 12 months?**	□ yes □ no
5.	**In the past 12 months, how often, on average, have you been kept awake at night by this itchy rash?**	□ never in the past 12 months □ less than one night per week □ one or more nights per week
6.	**Have you ever had eczema?**	□ yes □ no

Additional atopy questions

1. **From what age do you have symptoms suitable for asthma?**

   ….. year

   □ not applicable

2. **From what age do you have symptoms suitable for hayfever?**

   ….. year

   □ not applicable

3. **From what age do you have symptoms suitable for eczema?**

   ….. year

   □ not applicable

4. **Do you have a food allergy?**
   - □ yes, specify…
   - □ no
   - □ unknown

5. **What was your age at diagnosis of the food allergy?**

   ….. year

6. **Who diagnosed the food allergy?**
   - □ parent(s) and/or caregiver(s) based on complaints
   - □ a doctor based on complaints
   - □ a doctor based on a double-blind placebo-controlled food challenge
   - □ other, specify…
   - □ unknown

Appendix 7. Supplementary references

1. Odhiambo JA, Williams HC, Clayton TO, Robertson CF, Asher MI. Global variations in prevalence of eczema symptoms in children from ISAAC Phase Three. *J Allergy Clin Immunol* 2009;**124**(6):1251-1258 e1223.