

SUPPLEMENTARY TABLE II

A Blood counts, autoantibodies and functional assay findings in STING G207E carriers.

Patient	Control Range	IV.1	V.2	III.4	III.7	III.9	IV.6
Leukocytes (10 ⁶ /l)	3400-8200	4400	5100	4500	5900	5600	4200
Lymphocytes (10 ⁶ /l)	1300-3600	2240	1880	1600	2180	1860	1270
Monocytes (10 ⁶ /l)	200-800	190	110	540	500	220	100
Neutrophils (10 ⁶ /l)	1500-6700	1820	2840	2190	3040	3200	2580
Basophils (10 ⁶ /l)	0-100	20	10	10	10	10	0
Eosinophils (10 ⁶ /l)	30-440	120	270	180	120	300	100
B cells (CD19+) (10 ⁶ /l)	100-500	70	340	180	180	290	50
CD3+CD4+ (10 ⁶ /l)	300-1400	802	929	869	1065	1030	712
CD3+CD8+ (10 ⁶ /l)	200-1200	290	310	250	600	430	210
NK cells (CD3 ⁻ CD16 ⁺ 56 ⁺) (10 ⁶ /l)	90-600	50	180	180	280	120	200
Platelets (10 ⁶ /l)	150 000-360 000	107 000	254 000	229 000	279 000	274 000	173 000
ANA-ab (titer)	<320	<80	<80	<80	<80	<80	<80
ENA-ab	Negat	Negat	Negat	Negat	Negat	Negat	Negat
DNA-ab (IU/ml)	<10	<10	<10	NA	<10	<10	<10
ANCA	Negat	Negat	Negat	Negat	NA	Negat	NA
LupusAK	Negat	Negat	Negat	NA	Negat	Negat	NA
PLAb	Negat	Negat	NA	NA	Negat	NA	NA
KardAbG (U/ml)	<15	<4	<4	NA	<4	4	NA
IgE (kU/l)	0-110	290	4418	738	889	46	889
Complement	Classical, alternative, mannan-binding lectin pathway hemolytic activities	Normal	Normal	Normal	Normal	Normal	Normal
Lymphocyte proliferative responses to mitogens	Phytohemagglutinin, concanavalin A, pokeweed mitogen	Normal	Normal	NA	NA	Normal	NA

B Hematologic parameters of T- and B-cells of affected family members.

Patient	Control Range (%)	IV.1	V.2	III.4	III.7	III.9	IV.6
CD4-CD8-TCRab+	0,3-3,3	0.6	3.1	0.2	0.7	0.1	0.3
TCRgd+	1,9-11,7	0.4	2.8	2.5	3.0	1.1	2.0
CD3+CD4+ T cells	35.6–56.0	73.6	72.6	77.6	63	73.9	74.9
Naïve	CCR7 ⁺ CD45RA ⁺	62.8	69.3	71.3	59.9	61.3	76.1
TCM	CCR7 ⁺ CD45RA ⁻	30.2	19.6	20.9	27.8	24.8	14.6
TEM	CCR7 ⁻ CD45RA ⁻	6.3	7.9	6.7	11.3	11.3	8.5
TEMRA	CCR7 ⁻ CD45RA ⁺	0.7	1.7	1.0	1.0	2.6	0.8
Activated	HLADR ⁺ CD38 ⁻	4.5	4.3	8.4	7.4	7.0	3.6
	HLADR ⁻ CD38 ⁺	62.8	74.7	70.3	50.7	53.5	77.0
	HLADR ⁺ CD38 ⁺	3.3	3.9	5.0	3.9	2.0	1.9
RTE	CD45RA ⁺ CD62L ⁺ CD31 ⁺	28.8	43.2	28.6	16.7	19.0	24.5
Treg	CD25 ^{hi} CD127 ^{lo}	9.8	8.6	6.6	7.3	5.8	6.5
CD3+CD8+ T-cells	13.1–34.5	26.6	24.2	22.3	35.7	30.9	22.1
Naive	CCR7 ⁺ CD45RA ⁺	45.3	68.9	32.6	23.0	27.8	53.6
TCM	CCR7 ⁺ CD45RA ⁻	22.8	4.9	7.1	14.7	15.1	6.6
TEM	CCR7 ⁻ CD45RA ⁻	7.8	5.9	12.7	16.8	10.1	16.2
Temra	CCR7 ⁻ CD45RA ⁺	24.1	20.3	47.5	45.5	47.0	23.6
Activated	HLADR ⁺ CD38 ⁻	13.7	14.9	33.2	23.8	22.4	5.0
	HLADR ⁻ CD38 ⁺	19.3	50.1	15.1	15.1	11.6	39.1
	HLADR ⁺ CD38 ⁺	12.0	12.8	24.5	14.0	13.5	6.3
		290	4418	738	889	46	889
CD19+ B- cells	5-22	3	15	9	9	15	3
Naïve	CD27 ⁺ IgD ⁺	31.6	89.9	86	91.6	86.2	60.6
Activated	CD38 ^{low} CD21 ^{low}	12.4	6	7.5	2.8	1.5	9.1

Negat= negative, NA = not available. Values higher than control range are highlighted with red, whereas lower values are indicated with blue background.