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5	Letter to the Editor
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7	Defects in the CD19 complex predispose to glomerulonephritis as well as IgG1 subclass
8	deficiency.
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- 42 **Table E1.** Patients clinical and phenotypical characteristics. Subnormal values are in bold.
- 43 Immunizations were performed less than 5 years prior serum antibody detection.
- 44 Immunological work-up were performed before immunoglobulin substitution.

Characteristics	Control	Patient A	Patient B
Sex		F	F
Age at first symptoms - yr		5	13
Age at diagnosis - yr		11	29
Total lymphocytes - x10 ⁶ .L ⁻¹	1836 [1529-2350]	2745	1440
CD3+ T cells	1419 [1122-1774]	2141	1152
CD4+ T cells	912 [682-1153]	1125	590
CD4+CD45RA+CCR7+ naive T cells	363 [295-550]	532	175
CD4+CD25+CD127low T reg cells	67 [52-78]	71	na
CD8+ T cells	474 [356-616]	796	533
CD8+DR+ activated T cells	67 [43-105]	102	287
CD3-CD16+CD56+ NK cells	185 [115-270]	184	23
CD20+ B cells	202 [126-301]	300	61
Serum immunoglobulin level - mg/dl			
IgG	639-1349	230	493
IgA	70-312	125	350
IgM	56-352	35	135
IgG1	382-929	66	170
IgG2	242-700	144	306
IgG3	22-176	36	211
IgG4	2-86	<1	2
Blood group / Isohemagglutinin		O / Normal	
Serum antibodies			
polio		neg	pos
tetanus toxin		neg	pos
pertussis toxin		neg	low
pneumococcus		neg	na

45 Median values [IQR] of lymphocytes populations are presented for 50 controls ⁹; Serum

46 immunoglobulin ranges are from laboratory data; na, non available.

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- 48 **Table E2:** Lymphocyte subsets in *CD19* mutated patients. These experiments were realized
- 49 on whole blood.

		Units	Control (n=50)	Patient	APatient B
Lymphocyte	5	x10 ⁶ L ⁻¹	1836 [1529-2350]	2252	1568
B-cells	CD19	%	11 [9-14]	0.1	0.2
	CD20	%	11 [9-14]	13.3	3.9
CD81	CD20+CD81+/CD20	%	100 [98-100]	100	100
CD21	CD20+CD21+/CD20	%	97 [95-98]	99	92
Naives	CD20+CD27-IgD+/CD20	%	67 [54-76]	89	59
mzB	CD20+CD27+IgD+/CD20	%	15 [11-23]	8	22
smB	CD20+CD27+IgD-/CD20	%	15 [10-20]	2	12
CD21 ^{low}	CD20 ^{high} CD21-CD38-/CD20	%	2 [1-3]	0.4	2.9
Transitional	CD20+CD38++IgM++/CD20) %	2 [1-3]	0	0
Plasmablast	CD20+CD38++IgM-/CD20	%	0.9 [0.5-1]	0	0

50 Median values [IQR] are presented for 50 controls ⁹; smzB, marginal zone B cells; smB,

51 switched memory B cells.

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53 Figure Legends

- 54 **Figure E1.** Flow cytometric analysis of B cell subsets for patients A and B, patient A mother
- and a control. Experiments were performed on whole blood for patients and control and on
- 56 thawed PBMC for patient A mother.

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- 58 Figure E2. Flow cytometric analysis (a) and immunoblot (b) on EBV transformed cell lines
- 59 from patient A, patient B and control.

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62 Figure E1



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64 Figure E2