Otomo et al., http://www.jcb.org/cgi/content/full/jcb.201407077/DC1

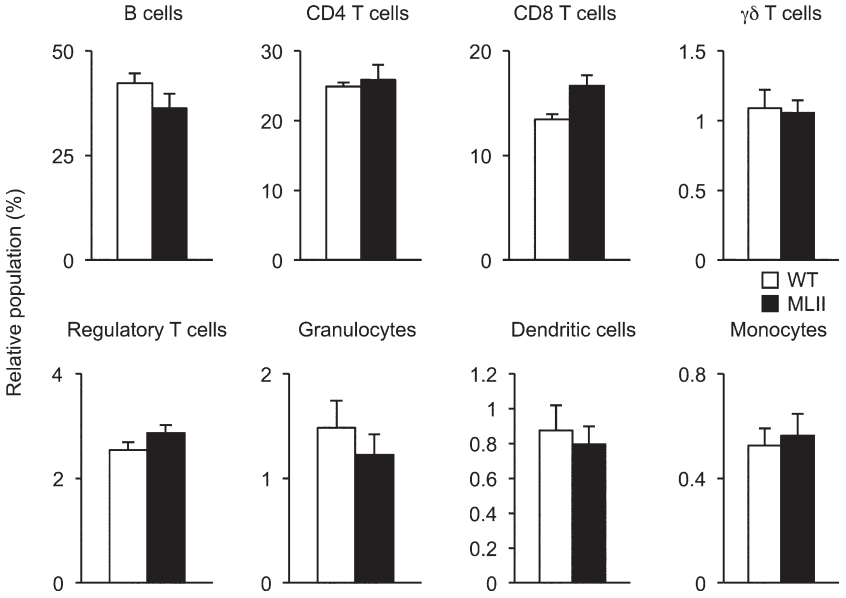

Figure S1. **Composition of immune cells in the spleen of MLII mice.** Immune cell populations in spleen from WT and MLII mice were analyzed by flow cytometry. Regulatory T cells are defined as $CD4^+Foxp3^+$ cells, granulocytes as $CD11b^+Gr1^{high}$ cells, DCs as $CD11c^+$MHC II^+ cells, and monocytes as $CD11b^+Ly6C^{high}Gr1^{low}$ cells. The data are expressed as the mean and SEM (n = 6–11, except regulatory T cells [n = 3]).

Table S1. **Flow cytometric analysis of lymphocytes in MLII patients**

Lymphocyte subset[a]	Patient 3	Patient 4	Reference range[b] (n = 33)
B cells	9	9	14–44
T cells	82	83	43–76
CD4/CD8 ratio	2.2	0.9	0.9–2.9
NK cells	6	7	4–23
B cell subset[c]	**Patient 3**	**Patient 4**	**Reference range[d] (n = 19)**
$CD27^-IgD^+$	93.0	ND	76.3–84.9
$CD27^+IgD^+$	3.7	ND	4.1–9.0
$CD27^+IgD^-$	1.6	ND	3.3–7.4
$CD27^-IgD^-$	1.6	ND	3.4–6.1

[a]Blood samples from MLII patients 3 and 4 analyzed by flow cytometry. The values represent percentage among lymphocytes with the exception of the ratio.
[b]Comans-Bitter et al., 1997.
[c]B cell subset analysis from MLII patient 3 revealed an enlarged naive $CD27^-IgD^+$ population and reduced IgD^- populations compared to the age-matched reference range, indicating impaired immunoglobulin class switching. The values represent percentage among B cells.
[d]Morbach et al., 2010.

Table S2. **Immune status of MLII patients**

Patient	*GNPTAB* mutations	Vaccination (response[a])	IVIG[b]	Ig levels (year/month)[c,d]
1	c.2089_2090insC (p.Leu697ProfsX51) c.3565C>T (p.Arg1189X)	Measles (−)	+	IgG 733, IgA 25, IgM 23 (1/8) IgG 742, IgA 28, IgM 18 (2/2)
2	c.3565C>T (p.Arg1189X) c.3565C>T (p.Arg1189X)	Measles (+), Mumps (+), Rubella (−), Varicella (−)	+	IgG 790, IgA 68, IgM 28 (1/5)
3	c.344_345delCA (p.Thr115AsnfsX5) c.1022delC (p.Pro341HisfsX18)	Diphtheria (−), Tetanus (+), Pertussis (−), Measles (−), Mumps (+), Rubella (−), Varicella (−)	−	IgG 571, IgA 37, IgM < 25 (2/5)
4	c.242G>T (P.Trp81Leu) c.242G>T (P.Trp81Leu)	Diphtheria (−), Tetanus (−), Pertussis (−), Measles (−), Mumps (−), Rubella (+), Hepatitis B (−)	−	IgG 540, IgA 81, IgM 32 (4/3)

[a]Antibody response: +, positive; −, negative/insufficient.
[b]IVIG, intravenous injections of immunoglobulins.
[c]Basal IgG, IgA, and IgM concentrations in serum (mg/dl) and the age of patients at the time of measurement: Values lower than age-matched controls are underlined.
[d]Stiehm and Fudenberg, 1966.

References

Comans-Bitter, W.M., R. de Groot, R. van den Beemd, H.J. Neijens, W.C. Hop, K. Groeneveld, H. Hooijkaas, and J.J. van Dongen. 1997. Immunophenotyping of blood lymphocytes in childhood. Reference values for lymphocyte subpopulations. *J. Pediatr.* 130:388–393. http://dx.doi.org/10.1016/S0022-3476(97)70200-2

Morbach, H., E.M. Eichhorn, J.G. Liese, and H.J. Girschick. 2010. Reference values for B cell subpopulations from infancy to adulthood. *Clin. Exp. Immunol.* 162:271–279. http://dx.doi.org/10.1111/j.1365-2249.2010.04206.x

Stiehm, E.R., and H.H. Fudenberg. 1966. Serum levels of immune globulins in health and disease: a survey. *Pediatrics*. 37:715–727.