Supplemental material to:

# Life-threatening primary varicella zoster virus infection with

# haemophagocytic lymphohistiocytosis-like disease in GATA2 haploinsufficiency accompanied by

# expansion of double negative T-lymphocytes

**Authors:** Seraina Prader1, MD, Matthias Felber1,2, MD, Benjamin Volkmer1, PhD, Johannes Trück1, MD, DPhil, Agnes Schwieger3, MD, Martin Theiler3,4, MD, Lisa Weibel3,4, MD, Sophie Hambleton5, MD, PhD, Katja Seipel6, PhD, Stefano Vavassori1, PhD, Jana Pachlopnik Schmid1,7, MD, PhD.

**Affiliations:**

1 Division of Immunology, University Children's Hospital Zurich, Zurich, Switzerland

2 Division of Stem Cell Transplantation, University Children's Hospital Zurich, Zurich, Switzerland

3 Department of Pediatric Dermatology, University Children's Hospital Zurich, Zurich, Switzerland

4 Department of Dermatology, University Hospital Zurich, Zurich, Switzerland

5 Institute of Cellular Medicine, International Centre for Life, Newcastle University, Newcastle upon Tyne, United Kingdom.

6 Department for Biomedical Research, University of Bern, Bern, Switzerland.

7 Pediatric Immunology, University of Zurich, Zurich, Switzerland

**Supplemental table 1. Leukocyte subpopulations before, during and after VZV infection in patient 1 and 2**

|  |  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- | --- |
|  |  | **Patient 1** |  | **empty** |  | **Patient 2** |  |
|  |  |  |  |  |  |  |  |
|  | **Before varicella infection**  **(- 3 months)** | **During varicella infection** | **After varicella infection**  **(+3 months)** |  | **Before varicella infection**  **(-3 weeks)** | **During varicella infection** | **After varicella infection**  **(+1 month)** |
| IgA (g/l) | 0.61 | nd | 0.58 |  | 1.21 | nd | 1.11 |
| IgM (g/l) | 1.13 | nd | 0.72 |  | 1.15 | nd | 1.29 |
| IgG (g/l) | 6.17 | 6.52 | 14.5 |  | 6.09 | nd | 7.67 |
| IgG1 (g/l) | nd | nd | 12.2 |  | nd | nd | 4.79 |
| IgG2 (g/l) | nd | nd | 1.17 |  | nd | nd | 1.12 |
| IgG3 (g/l) | nd | nd | 1.34 |  | nd | nd | 0.29 |
| IgG4 (g/l) | nd | nd | <0.07 |  | nd | nd | <0.07 |
| Tetanus IgG (U/ml) | 577 | nd | 377 |  | nd | nd | nd |
| HiB IgG (µg/ml) | 3.14 | nd | 5.14 |  | nd | nd | nd |
| Leukocytes (x 109/l) | 2.8 | 2.67 | 6.78 |  | 2.39 | 1.3 | 1.63 |
| Lymphocytes (x 109/l) | 1.31 | 1.34 | 2.32 |  | 0.88 | 0.38 | 1.12 |
| TCR alpha/beta (x 109/l) | 1.17 | 1.31 | 2 |  | 0.55 | nd | 0.75 |
| TCR gamma/delta  (x 109/l) | 0.01 | 0 | 0.05 |  | 0.28 | nd | 0.31 |
| DNT\* | 13.3% | 1.6% | 11.2% |  | 20.9% | 22.5% | 20.4% |
| CD4+ (x 109/l) | 0.68 | 0.86 | 0.83 |  | 0.24 | 0.1 | 0.34 |
| CD4+CD45RA+ (x 109/l) | 0.46 | 0.61 | 0.64 |  | 0.15 | nd | 0.22 |
| CD31+CD45RA/CD4+ (x 109/l) | 0.22 | 0.31 | 0.35 |  | 0.06 | nd | 0.07 |
| CD8+ (x 109/l) | 0.48 | 0.46 | 1.18 |  | 0.46 | 0.21 | 0.55 |
| CD19+ (x 109/l) | 0.05 | 0.01 | 0 |  | 0.03 | 0.02 | 0.02 |
| CD20+ (x 109/l) | 0.04 | 0 | 0 |  | 0.03 | 0.01 | 0.01 |
| CD16+CD56+ (x 109/l) | 0.07 | 0 | 0.26 |  | 0.01 | 0 | 0.01 |
| CD14+ (x 109/l) | 0.04 | 0 | 0 |  | 0.02 | nd | nd |

\*DNT: % of double negative T cells

### Supplemental table 2. Antibodies used for Mass Cytometry

|  |  |
| --- | --- |
| **Target** | **Clone** |
| CD196/CCR6 | G034E3 |
| CD19 | HIB19 |
| CD127 | A019D5 |
| CD11b | ICRF44 |
| CD4 | RPA-T4 |
| CD8a | RPA-T8 |
| CD16 | 3G8 |
| CD25 (IL-2R) | 2A3 |
| CD138 | DL-101 |
| CD192 (CCR2) | K036C2 |
| CD45 | HI30 |
| CD279/PD-1 | EH12.2H7 |
| CD195/CCR5 | NP-6G4 |
| CD194/CCR4 | 205410 |
| CCR7 | G043H7 |
| CD28 | CD28.2 |
| CD69 | FN50 |
| CD294/CRTH2 | BM16 |
| CCR10 | 314305 |
| CD27 | O323 |
| CD45RA | HI100 |
| CD3 | UCHT1 |
| CXCR5 | RF8B2 |
| CD38 | HIT2 |
| HLA-DR | L243 |
| CD14 | M5E2 |
| CD56 | NCAM16.2 |
| CD45 | HI30 |