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

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**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 |