**Table e-2.** Long-term immunologic follow-up.

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| --- | --- | --- | --- | --- |
| **Patients on follow-up** | **N=20**  **Baseline** | **N=14\*\***  **12 months after RTX** | **N=8\*\*\***  **18 months after RTX** | **N=6\*\*\*\***  **24 months after RTX** |
| Number of patients with low % of B cells | 2/17 (11.8%) | 9/14 (64.3%) | 4/8 (40%) | 3/6 (50%) |
| Number of patients with %B cells 0 - 2% | 0/17 | 3/14 (35.7%) | 2/8 (25%) | 2/6 (33.3%) |
| Complete depletion of B cells | 0/20 | 2/14 (14,3%) | 0/8 | 0/6 |
| Number of patients with dysgammaglobulinemia | 1\*/20 | 5/14 (35.7%) | 3/8 (37.5%) | 2/6 (33.3%) |
| Isolated hypo-IgG | 1\*/20 | 1/14 (7.1%) | 0/8 | 0/6 |
| Isolated hypo-IgA | 0/20 | 1/14 (7.1%) | 1/8 (12.5%) | 0/6 |
| Isolated hypo-IgM | 0/20 | 0/14 | 0/8 | 0/6 |
| Hypo-IgG and IgA | 0/20 | 1/14 (7.1%) | 1/8 (12.5%) | 1/6 (16.6%) |
| Hypo-IgA and IgM | 0/20 | 1/14 (7.1%) | 0/8 | 0/6 |
| Hypo IgG and IgM | 0/20 | 1/14 (7.1%) | 1/8 (12.5%) | 1/6 (16.6%) |
| Number of patients requiring IVIG post-RTX | 0 | 1/14 (7.1%) | 0 | 0 |
| Number of patients with high % naive B cells | 1/20 (5,0%) | 5/11 (45.5%) | 1/4 (25%) | 1/4 (25%) |
| Number of patients with low % switched-memory B cells | 2/20 (10%) | 3/9 (33.3%) | 2/4 (50%) | 0/4 |
| Number of patients with plasmablasts below 1% | 7/20 (35.0%) | 3/11 (27.3%) | 1/4 (25%) | 3/4 (75%) |
| Infections post RTX | 0/20 | 1 | 0 | 0 |

IVIG: intravenous Immunoglobulin

\* At onset and during the follow-up, a patient with nephrotic syndrome had hypo IgG, but as hypo IgG correlated with hypoalbuminemia, it was not considered pathological, but was not included for the IgG kinetics analysis.

\*\* 6/20 children were excluded for the 12 month follow-up analysis as 3 had RTX-retreatment, 2 had <12 months post-RTX follow-up, and in one the data was missing for the 12 month timepoint, Figure e1).

\*\*\*for the 18 months follow-up additional 4 patients were excluded as they had received RTX, and 3 patients were excluded for the analysis as they had previously reached complete immune recovery and no additional follow-up was performed as per protocol (Table 1)..

\*\*\*\*for the 24 months follow-up additional 2 patients were excluded as they had previously reached complete immune recovery an no additional follow-up was performed as per protocol (Table 1)..