**Table e-1.** Main characteristics of the cohort receiving a single cycle of Rituximab

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| --- | --- |
| **Patients** | **N=20** |
| Gender | 12 (60%) female / 8 (40%) male |
| Ethnicity | 17 (85%) Caucasians  1 (5%) Asian  2 (10%) Arabs |
| Consanguinity | 1 (5%) |
| Family history of autoimmune diseases | 5 (25%) |
| Underlying immune-mediated disorder | 9 (45%) neuro-immunological diseases1  1 (4.5%) dermatological diseases2  8 (36.4%) nephrological diseases3  2 (9.1%) rheumatic disease4 |
| Median age (years) at disease onset  (median / IQR) | 8.5 (3-13.75) |
| Median age (years) at first RTX infusion  (median/ IQR) | 12.8 (6.6-15.5) |
| Median disease duration before  first RTX (years) | 1.9 (0.4-4.5) |
| Number of patients with hypogammaglobulinemia (hypo IgG) prior to RTX | 1 (5%)5  (nephrotic syndrome) |
| Number of patients with low % of B-lymphocytes prior to RTX6 | 2 (10%) |
| Median follow-up after RTX treatment (months) | 12.6 (10.2-24) |
| Immunosuppressive treatment before RTX | 20 (100%):  steroids alone 9/20;  steroids + second-line immunosuppressants\*: 11/20 |
| Immunosuppressive treatment during RTX | 11 (55%):  steroids alone 5/11;  steroids + second-line immunosuppressants\*\*: 6/11 |

13 anti-NMDAR encephalitis, 6 recurrent anti-MOG-associated inflammatory central nervous system disease; 2Paraneoplastic pemphigus; 33 Nephrotic syndrome with chronic renal failure, 2 ANCA glomerulonephritis and vasculitis, and 3 acute antibody-mediated rejection of kidney transplant; 4Both juvenile idiopathic arthritis; 5One patient with nephrotic syndrome had hypo IgG due to severe proteinuria but as hypo IgG correlated with hypoalbuminemia and disease activity this was not considered pathological or related to RTX – effect.

\*second-line immunosuppressants included: mycophenolate, azathioprine, methotrexate, cyclosporine A, cyclophosphamide , tacrolimus.

\*\*second-line immunosupressants included: mycophenolate, azathioprine, methotrexate, cyclosporine A, tacrolimus.

Abbreviations: Ig: immunoglobulins; NMDAR: N-methyl-D-asparatate receptor; MOG: myelin-oligodendrocyte glycoprotein; RTX: rituximab.