**Supplementary materials**

**Pediatric Primary Immune Regulation Disorder on IL-10 -IL-10R Axis: A systematic review of 284 patients**

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**Figure S1- Initial diagnosis of patients with IL-10 and/or IL-10R deficiency.**



**Figure S2-** Kaplan-Meier survival curves of all HSCT transplanted and transplanted cases **(A),** patients with IL-10R deficiency **(B),** patients with IL-10RA deficiency **(C),** patients with IL-10RB deficiency **(D)**. Transplantation was associated with a higher survival rate in all (**A-D)** curves.

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**Figure S3-** Kaplan-Meier survival curves of cases with available data who underwent HSCT.

**Table S1– Therapeutic data of patients with IL-10/IL-10R deficiencies.**

|  |  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- | --- |
| **Parameters** | **Total** | **IL-10** | **IL-10R** | ***P- value*** | **IL-10RA** | **IL-10RB** | ***P- value*** |
| **Transplantation, n, % (n=187)** | 56 (29.9) | 2 (50) | 54 (29.5) | 0.638 | 37 (28.5) | 15 (30) | 0.838 |
| **Transplantation age, y, median (IQR), (n=157)** | 2.0 (1.0-6.0) | 2.5 (1.2- ) | 2.0 (0.9-6.3) | 1.000 | 1.7 (0.8-5.8) | 3.0 (1.0-7.5) | 0.330 |
| **Follow up after transplantation, y, median (IQR), (n=157)** | 1.0 (0.5-1.5) | - | 1.0 (0.5-1.5) | **-** | 0.5 (0.3-1.2) | 1.0 (1.0-2.0) | **0.015** |
| **Neutrophil engraftment, d, median (IQR), (n=157)** | 21.0 (13.0-23.2) | 0 | 21.0 (13.0-23.2) | *-* | 21.5 (14.5-28.5) | 12.0 (11.0-18.5) | 0.058 |
| **Thrombocyte engraftment, d, median (IQR), (n=157)** | 26.0 (19.0-29.0) | 0 | 26.0 (19.0-29.0) | *-* | 27.5 (25.2-30.7) | 14.0 (9.2-22.5) | **0.002** |
| **Immunosuppressive therapy, n, % (n=184)** | 160 (87) | 8 (100) | 152 (86.4) | 0.097 | 104 (84.6) | 46 (90.2) | 0.326 |
| **Steroid therapy, n, % (n=163)** | 107 (65.6) | 7 (100) | 100 (64.1) | 0.599 | 61 (55) | 37 (86) | **<0.001** |
| **Surgery, n, % (n=174)** | 107 (61.5) | 3 (42.9) | 106 (57.6) | 0.465 | 64 (49.6) | 38 (74.5) | **0.002** |
| *y; year, d; day, n; number**The median is shown [with 25th and 75th percentiles].**\* p-value is statistically significant <0.05* |