**JPED-D-21-00106\_Supplementary Material**

**Supplementary Table 1 - Clinical and laboratory data of children with hypogammaglobulinemia (n = 22) pre and post 12 months’ intravenous immunoglobulin (IVIG) therapy.**

|  |  |  |  |
| --- | --- | --- | --- |
|  | **Pre-IVIG** | **Post-IVIG** | ***P* value** |
| Number of HA per patient per year  | 1.5 (0-30) | 0 (0-5) | <0.001 |
| LOS per patient per year (days) | 14 (0-167) | 0 (0-55) | <0.001 |
| LOS in PICU per patient per year (days) | 0 (0-105) | zero | <0.002 |
| Pneumonia episodes per patient per year | 0.7 (0-6) | 0 (0-2) | <0.001 |
| URTI episodes per patient per year | 0 (0-1.13) | 0 (0-1) | 0.09 |
| IgG (mg/dL) | 268.5 (78-611) | 706 (478-1200) | <0.001 |
| IgM (mg/dL) | 22.8 (9.8-200) | 45.1 (5-204) | 0.99 |
| IgA (mg/dL) | 33 (1-132) | 42.5 (0-138) | 0.72 |
| CD19/μL | 1203(3-1510) |  |  |

HA, Hospital admission; LOS, length of stay; PICU, Pediatric intensive care unit; URTI, Upper respiratory tract infection. Values are expressed in median (range). Statistical significance when p < 0.05. Wilcoxon test.

**Supplementary Table 2- Demographic and main laboratory results of the cohort of patients with Inborn errors of immunity (n = 45).**

|  |  |  |  |  |  |  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- |
| **Patient** | **Gender** | **Age at diagnosis****(months)** | **Diagnosis** | **IVIG dose (mg/kg)** | **Antibiotics** | **IgG-Pre (mg/dL)** | **IgG-Post****(mg/dL)** | **IgM-Pre****(mg/dL)** | **IgM-Post****(mg/dL)** | **IgA-Pre****(mg/dL)** | **IgA-Post****(mg/dL)** | **CD19-Pre** |
| 1 | F | 15 | THI | 400 | No | 382 | 675 | 28,5 | 36,6 | 25,5 | 27 | 897 |
| 2 | M | 119 | CVID | 519 | No | 511 | 1030 | 111,0 | 204 | 23 | 25,3 | 169 |
| 3 | F | 121 | HIES+SAD | 511 | Yes | 1318 | 2467 | 177 | 129 | 111 | 122 | 377 |
| 4 | M | 47 | PNP def. | 514 | No | 1260 | 676 | 234 | 44,9 | 160,8 | 51,7 | 22 |
| 5 | M | 24 | XLA | 500 | No | 141 | 631 | 15 | 17 | 24,4 | 25,5 | 3 |
| 6 | M | 95 | CVID | 725 | No | 78 | 639 | 9,8 | 5 | 6 | 0 | 335 |
| **7** | M | 61 | SAD | 417 | No | 1360 | 1540 | 90,1 | 108 | 36 | 206 | 630 |
| 8 | F | 8 | SAD | 500 | Yes | 1460 | 883 | 202 | 150,6 | 180 | 119 | 525 |
| 9 | M | 7 | THI | 400 | No | 244 | 893 | 20 | 51,5 | 22,8 | 29,9 | 1303 |
| 10 | M | 116 | SAD | 580 | No | 1460 | 1391 | 119 | 134 | 134 | 142 | 275 |
| 11 | M | 156 | HIES+SAD | 600 | Yes | 1470 | 1690,0 | 202,0 | 134,0 | 147 | 169,0 | 390 |
| 12 | M | 31 | OH | 500 | Yes | 212 | 984 | 58 | 71,3 | 28 | 54,9 | 1091 |
| 13 | M | 5 | THI | 400 | No | 135 | 697 | 56,5 | 40,2 | 26 | 26 | 1029 |
| 14 | F | 5 | OH | 657 | No | 204 | 690 | 20 | 8,1 | 23,3 | 9 | 128 |
| 15 | F | 104 | SAD | 545 | No | 1474 | 1300 | 99 | 128 | 355 | 373 | 91 |
| 16 | M | 117 | CVID | 400 | No | 611 | 1073 | 92 | 145,5 | 34 | 46 | 225 |
| 17 | M | 48 | OH | 458 | No | 350 | 697 | 129 | 67,8 | 64 | 55 | 284 |
| 18 | M | 135 | SAD | 400 | No | 1200 | 1080 | 180 | 79.2 | 133 | 167 | 149 |
| 19 | F | 102 | CVID | 500 | No | 257 | 673 | 19 | 37 | 26 | 3 | 272 |
| 20 | F | 15 | SAD | 610 | Yes | 1200 | 1393 | 180 | 85,7 | 133 | 31 | 149 |
| 21 | M | 13 | SAD | 500 | No | 840 | 1081 | 134 | 108 | 182 | 242 | 1406 |
| 22 | M | 53 | XLA | 598 | No | 137 | 734 | 50,1 | 12,4 | 5 | 0 | 10 |
| 23 | M | 167 | SAD | 667 | Yes | 921 | 1195 | 93,1 | 71 | 282 | 249 | 534 |
| 24 | M | 75 | HIES+SAD | 600 | No | 1120 | 1440 | 42 | 45,8 | 197 | 268 | 314 |
| 25 | M | 24 | OH | 500 | No | 295 | 909 | 107 | 30 | 25 | 19 | 506 |
| 26 | F | 38 | SAD | 625 | Yes | 1022 | 908,0 | 134,3 | 152,0 | 293 | 84,0 | 348 |
| 27 | F | 77 | OH | 540 | No | 518 | 941 | 200,0 | 169 | 50 | 45 | 493 |
| 28 | M | 71 | SAD | 512 | No | 1170 | 1730 | 287,0 | 217 | 186 | 243 | 75 |
| 29 | M | 76 | SAD | 598 | No | 1279 | 1520 | 252 | 38 | 404 | 509 | 149 |
| 30 | M | 126 | AT | 555 | No | 1163 | 1418 | 216,0 | 290 | 3 | 6 | 117 |
| 31 | F | 6 | CGD+Hypogamma | 568 | Yes | 137 | 1246 | 19,4 | 90,6 | 24,5 | 62 | 3296 |
| 32 | F | 73 | SAD | 500 | No | 846 | 939 | 87 | 57,4 | 80,9 | 85 | 243 |
| 33 | F | 156 | CVID | 640 | No | 204 | 478 | 24 | 14 | 20 | 15 | 325 |
| 34 | M | 180 | OH | 690 | No | 85 | 826 | 66,0 | 82 | 110 | 138 | 729 |
| 35 | M | 20 | THI | 545 | No | 286 | 679 | 62,1 | 781 | 33 | 33 | 743 |
| 36 | M | 30 | XLA | 500 | No | 78,9 | 717 | 17,3 | 17,9 | 25 | 26 | 10 |
| 37 | M | 11 | THI | 450 | No | 274 | 715 | 51 | 49,3 | 25,8 | 57,5 | 1410 |
| 38 | F | 3 | OH | 537 | No | 157 | 773 | 72 | 20,8 | 7 | 28 | 299 |
| 39 | F | 154 | Hyper IgM | 800 | Yes | 346 | 1051 | 1482 | 1503 | 1 | 1 | 1406 |
| 40 | M | 119 | OH | 500 | No | 413 | 792 | 149 | 146 | 132 | 124 | 50 |
| 41 | M | 6 | THI | 588 | No | 155 | 737 | 17 | 53,7 | 40,7 | 58,1 | 1510 |
| 42 | F | 112 | HIES+SAD | 479 | Yes | 647 | 977 | 90 | 70 | 414 | 334 | 148 |
| 43 | F | 204 | SAD | 500 | No | 1286 | 1500 | 166,0 | 67 | 126 | 137 | 75 |
| 44 | M | 88 | XLP | 517 | No | 322 | 745,0 | 46,0 | 79,0 | 33 | 26,0 | 246 |
| 45 | M | 125 | SAD | 625 | Yes | 709 | 1207,0 | 105,0 | 119,0 | 32 | 127,0 | 384 |

THI, Transient Hypogammaglobulinemia of infancy; CVID, Common variable immunodeficiency; OH, Other hypogammaglobulinemia; SAD, Specific antibody deficiency; HIES, Hyper IgE syndrome; XLA, X-linked agammaglobulinemia; XLP, X-linked proliferative disease.

|  |  |  |
| --- | --- | --- |
|   | Antibiotic use  |   |
|  | No (n = 34) | Yes (n = 11) | p |
|   | Median | IQR | Median | IQR |
| IgG ( mg/dL) | 809.0 | 441.0 | 1195.0 | 416.0 | **0.02** |
| IgM (mg/dL) | 55.5 | 101.9 | 119.0 | 79.3 | **0.03** |
| IgA (mg/dL) | 45.5 | 113.6 | 119.0 | 333.0 | 0.16 |
| Leucocytes/ μL | 6500.0 | 2824.3 | 5600.0 | 3200.0 | 0.29 |
| Lymphocytes/ μL  | 2850.0 | 2437.8 | 2240.0 | 2100.0 | 0.41 |
| CD3/ μL | 1236.0 | 1975.8 | 1484.0 | 1803.0 | 0.65 |
| CD4/μL | 905.0 | 730.0 | 960.5 | 1003.0 | 0.89 |
| CD8/μL | 744.0 | 889.0 | 557.5 | 333.0 | 0.19 |
| CD19/μL | 254.0 | 385.8 | 422.5 | 607.0 | 0.11 |
| Hospital admission (number) | 0.0 | 0.00 | 0.0 | 3.0 | 0.34 |
| LOS in general nursery (days) | 0.0 | 0.30 | 0.0 | 24.0 | 0.38 |
| LOS in PICU (days) | 0.0 | 0.00 | 0.0 | 0.0 | 1.00 |
| Pneumonia (number) | 0.0 | 0.00 | 0.0 | 0.0 | 0.63 |
| URTI (number) | 0.0 | 0.00 | 0.0 | 1.0 | 0.07 |
|   | Immunosuppressive medication  | p |
|  | No (n = 41) | Yes (n = 4) |
| IgG (mg/dL) | 939.0 | 548.0 | 1036.0 | 951.0 | 0.86 |
| IgM (mg/dL) | 79.0 | 1498.0 | 52.9 | 86.2 | 0.30 |
| IgA(mg/dL) | 54.9 | 114.0 | 91.00 | 388.6 | 0.57 |
| Leucocytes/μL | 6400.0 | 3048.5 | 5850.0 | 4493.0 | 0.40 |
| Lymphocytes/μL | 2900.0 | 2326.5 | 1503.0 | 1340.0 | 0.07 |
| CD3/μL | 1267.0 | 1896.0 | 1165.0 | 1683.8 | 0.52 |
| CD4/μL | 931.5 | 732.5 | 912.0 | 663.0 | 0.59 |
| CD8/μL | 635.5 | 815.3 | 793.0 | 417.0 | 0.87 |
| CD19/μL | 381.0 | 570.0 | 208.0 | 94.0 | 0.31 |
| Hospital admission (number) | 0.0 | 0.5 | 0.0 | 0.8 | 0.95 |
| LOS in general nursery (days) | 0.0 | 3.0 | 0.0 | 9.8 | 1.00 |
| LOS in PICU (days) | 0.0 | 0.0 | 0.0 | 0.0 | 1.00 |
| Pneumonia (number) | 0.0 | 0.0 | 0.0 | 0.0 | 0.71 |
| URTI (number) | 0.0 | 0.0 | 0.0 | 0.0 | 0.77 |

 **Supplementary Table 3 - Comparative analysis of patients with Inborn Errors of Immunity under intravenous immunoglobulin antibiotics with and without associated medications (antibiotics/immunosuppressive medication). Hospital admission data and laboratory data were studied (n = 45).**

LOS, Length of stay; PICU, Pediatric intensive care unit; URTI, Upper respiratory tract infection. Statistical significance when p < 0.05. U Mann-Whitney test.