Supplemental Table 1:. Relevant Immunologic Findings for Patient’s Daughter

|  |  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- | --- |
| Laboratory Parameter | Reference Values | 4 years | 5 years | 6 years | 7 years | 8 years | 9 years |
| WBC (x 10^3/µL) | 4-5 y.o 5.0-14.56-7 y.o. 5.0-14.58-9 y.o. 5.0-14.5 | 2.6 | 5.78 | 4.53 | 3.79 | 4.8 | 5.11 |
| RBC (x 10^6/µL) | 4-5 y.o. 3.9-5.36-7 y.o. 3.9-5.38-9 y.o. 4.0-5.2 | 4.43 | 5.17 | 5.15 | 5.01 | 5.46 | 5.38 |
| Hgb (g/dL) | 4-5 y.o. 11.5-14.56-7 y.o. 11.5-14.58-9 y.o. 11.5-15.5 | 12.2 | 14.3 | 14.2 | 13.4 | 14.7 | 13.9 |
| HCT (%) | 4-5 y.o 34.0-40.06-7 y.o. 34.0-40.08-9 y.o. 35.0-45.0 | 33.8 | 41.1 | 40.1 | 39.4 | 43.2 | 41.4 |
| Platelet (x 10^3/µL) | 4-5 y.o. 135-3616-7 y.o. 150-4508-9 y.o 150-450 | 10 | 82 | 122 | 168 | 195 | 196 |
| ANC (x 10^3/µL) | 4-5 y.o 1.9-10.36-7 y.o. 1.5-8.08-9 y.o. 1.5-8.0 | 0.12 | 1.51+ | 1.61 | 1.01 | 1.12 | 1.68 |
| IgA (MG/DL) | 4-5 y.o. 25-1546-7 y.o. 32-1918-9 y.o. 42-223 | 13 | TNP | 9.5 | 7.4 | 7.6 | <6.0 |
| IgG (MG/DL) | 4-5 y.o 463-1,2366-7 y.o. 635-1,2848-9 y.o. 610-1,577 | 366 | 498\* | 511\* | 421\* | 1,110\* | 1,010\* |
| IgM (MG/DL) | 4-5 y.o. 43-1966-7 y.o. 44-1908-9 y.o. 48-222 | 18 | TNP | 11.1 | 10.3 | 13.4 | 19.9 |
| Absolute Lymph Count (10^3/µL) | 10^3/µL | 2,880 | 2,784 | 2,360 | 1,672 | 1,968 | 1,887 |
| CD3+T Cell Percent (%)  | 4-5 y.o. 58-746-7 y.o. 56-768-9 y.o. 56-75 | 68 | 70 | 73 | 72.4 | 72.6 | 71.3 |
| CD3+T Cell Number (10^3/µL) | 4-5 y.o. 1,656-3,8416-7 y.o. 991-2,9978-9 y.o. 1,200-2,600 | 1,958 | 1,949 | 1,723 | 1,211 | 1,429 | 1,345 |
| CD3+CD4+Percent (%) | 4-5 y.o. 28-476-7 y.o. 25-488-9 y.o. 31-47 | 35.6 | 40.3 | 42.3 | 42.6 | 41.4 | 38.7 |
| CD3+CD4+Number (10^3/µL) | 4-5 y.o. 871-2,3796-7 y.o. 635-1,6208-9 y.o. 650-1,500 | 1,025 | 1,122 | 998 | 712 | 815 | 730 |
| CD3+CD8+Percent (%) | 4-5 y.o. 16-326-7 y.o. 16-438-9 y.o. 18-35 | 25.9 | 22.9 | 22.9 | 21.6 | 22.7 | 25.5 |
| CD3+CD8+Number (10^3/µL)  | 4-5 y.o 518-1,4336-7 y.o. 293-1,2218-9 y.o. 370-1,100 | 746 | 638 | 540 | 361 | 447 | 481 |
| CD3+/CD4-/CD8-Percent (%) |  |  |  |  | 7.0 | 7.9 | 5.2 |
| CD19+B Cell Percent (%) | 4-5 y.o. 13-316-7 y.o. 11-288-9 y.o. 13-27 | 23 | 22 | 19 | 16.8 | 19.1 | 17.6 |
| CD19+B Cell Number (10^3/µL) | 4-5 y.o. 421-1,3976-7 y.o. 249-8658-9 y.o. 270-860 | 662 | 612 | 448 | 281 | 376 | 332 |
| CD3-CD56CD 16+Percent (%) | 4-5 y.o. 3-196-7 y.o. 5-218-9 y.o. 4-17 | 8.0 | 7.0 | 7.0 | 10.1 | 8.1 | 11.0 |
| CD3-CD56CD 16+Number (10^3/µL) | 4-5 y.o. 123-7856-7 y.o. 128-4748-9 y.o. 100-480 | 230 | 195 | 165 | 169 | 159 | 208 |
| CD4 to CD8 Ratio | 4-5 y.o. 0.7-2.96-7 y.o. 0.7-2.68-9 y.o. 0.7-2.6 | 1.37 | 1.76 | 1.85 | 1.97 | 1.82 | 1.52 |
| CD3+CD56CD16+Percent (%) | 4-5 y.o. 0-106-7 y.o. 0-108-9 y.o. 0.5-23 | 1.0 | 1.0 | 1.0 | 1.8 | 2.3 | 2.6 |
| CD3+CD56CD16+Number (10^3/µL) | 4-5 y.o. 0-2006-7 y.o. 0-2008-9 y.o. 0-766 | 29 | 28 | 24 | 30 | 45 | 49 |
| CD19+CD27-IgM+IgD+Percent (%) | 4-5 y.o. 31-916-7 y.o. 43-898-9 y.o. 43-89 | 81.1 | TNP | TNP | 91.5 | 87.3 | 87.1 |
| CD19+CD27-IgM+IGD+Number (10^3/μL) | 4-5 y.o. 12-3366-7 y.o. 3-3838-9 y.o. 3-383 | 464 | TNP | TNP | 243 | 333 | 337 |
| CD19+CD27+Percent (%) | 4-5 y.o. 0-586-7 y.o. 2-368-9 y.o. 2-36 | 7.9 | TNP | TNP | 5.8 | 5.7 | 11.3 |
| CD19+CD27+Number (10^3/μL) | 4-5 y.o. 0-2046-7 y.o. 2-1228-9 y.o. 2-122 | 45 | TNP | TNP | 15 | 22 | 44 |
| CD19+CD27+IgM+IgD+Percent (%) | 4-5 y.o. 0-406-7 y.o. 0-198-9 y.o. 0-19 | 5.5 | TNP | TNP | 5.0 | 5.4 | 10.8 |
| CD19+CD27+IgM+IgD+Number (10^3/μL) | 4-5 y.o. 0-1406-7 y.o. 0-508-9 y.o. 0-50 | 31 | TNP | TNP | 13 | 21 | 42 |
| CD19+CD27+IgM+IgD-Percent (%) | 4-5 y.o. 0-46-7 y.o. 0-58-9 y.o. 0-5 | 0.1 | TNP | TNP | 0 | 0 | 0.1 |
| CD19+CD27+IgM+IgD-Number (10^3/μL) | 4-5 y.o. 0-106-7 y.o. 0-88-9 y.o. 0-8 | 1.0 | TNP | TNP | 0 | 0 | 0 |
| CD19+CD27+IgM-IgD-Percent (%) | 4-5 y.o. 0-306-7 y.o. 0-228-9 y.o. 0-22 | 2.3 | TNP | TNP | 0.7 | 0.2 | 0.3 |
| CD19+CD27+IgM-IgD-Number (10^3/μL) | 4-5 y.o. 0-946-7 y.o. 0-748-9 y.o. 0-74 | 13 | TNP | TNP | 2.0 | 1.0 | 1.0 |
| CD19+CD20+Percent (%) | 4-5 y.o. 97-1006-7 y.o. 96-1008-9 y.o. 96-100 | 99.3 | TNP | TNP | 99.8 | 98.5 | 99.9 |
| CD19+CD20+Number (10^3/μL) | 4-5 y.o. 42-5246-7 y.o. 22-5468-9 y.o. 22-546 | 568 | TNP | TNP | 265 | 376 | 386  |
| CD19+CD38+Bright IgM+Bright Percent (%) | 4-5 y.o. 0-76-7 y.o. 0-68-9 y.o. 0-6 | 5.9 | TNP | TNP | 14.3 | 7.5 | 9.1 |
| CD19+CD38+Bright IgM+Bright Number (10^3/μL) | 4-5 y.o. 0-216-7 y.o. 0-188-9 y.o. 0-18 | 34 | TNP | TNP | 38 | 29 | 35 |
| CD19+CD38+Bright IgM- Percent (%) | 4-5 y.o. 0-36-7 y.o. 0-38-9 y.o. 0-3 | 0.9 | TNP | TNP | 1.1 | 0.5 | 0.1 |
| CD19+CD38+Bright IgM- Number (10^3/μL) | 4-5 y.o. 0-66-7 y.o. 0-78-9 y.o. 0-7 | 5.0 | TNP | TNP | 3.0 | 2.0 | 0 |
| CD19+CD38+Bright Percent (%) | 4-5 y.o. 0-166-7 y.o. 0-148-9 y.o. 0-14 | 15.4 | TNP | TNP | 21.7 | 17.1 | 16.5 |
| CD19+CD38+Bright Number (10^3/μL) | 4-5 y.o. 0-506-7 y.o. 1-398-9 y.o. 1-39 | 88 | TNP | TNP | 58 | 65 | 64 |
| CD19+CD21+ Percent  (%) | 4-5 y.o. 88-1006-7 y.o. 86-1008-9 y.o. 86-100 | 94.2 | TNP | TNP | 91.9 | 86.5 | 95.9 |
| CD19+CD21+ Number (10^3/μL) | 4-5 y.o. 35-5086-7 y.o. 17-5228-9 y.o. 17-522 | 539 | TNP | TNP | 244 | 330 | 371 |
| CD19+CD38+CD21+ Percent (%) | 4-5 y.o. 49-1006-7 y.o. 57-968-9 y.o. 57-96 | 90.2 | TNP | TNP | 88 | 87.5 | 87.2 |
| CD19+CD38+CD21+ Number (10^3/μL) | 4-5 y.o. 16-4316-7 y.o. 16-4258-9 y.o. 16-425 | 516 | TNP | TNP | 234 | 334 | 337 |
| CD19+CD38-/LowCD21-/Low Percent (%) | 4-5 y.o. 0-86-7 y.o. 0-78-9 y.o. 0-7 | 1.6 | TNP | TNP | 1.7 | 1.7 | 1.7 |
| CD19+CD38-/LowCD21-/Low Number (10^3/μL) | 4-5 y.o. 0-196-7 y.o. 0-218-9 y.o. 0-21 | 9.0 | TNP | TNP | 5.0 | 6.0 | 7.0 |
| Proliferation to Mitogens & Antigens | NA | Normal | TNP | Normal | Normal | Normal | TNP |

\*IgG obtained while patient’s daughter was receiving IgG replacement therapy

+ANC obtained while patient’s daughter was receiving G-CSF therapy

 TNP: test not performed