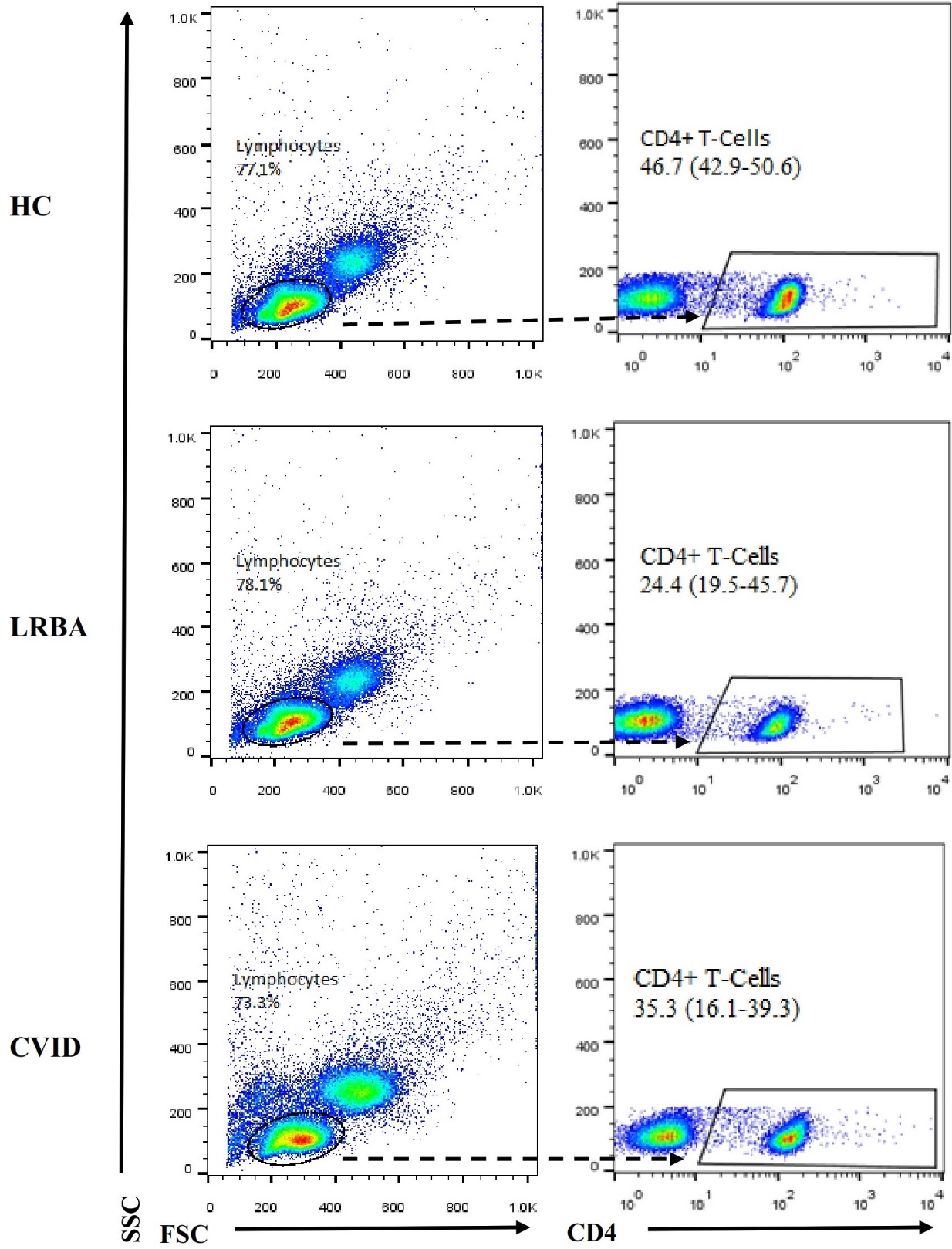
**Supplementary materials**

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| **Table S1-Oligonucleotide sequences of primers used in this study for quantitative real time PCR** | | |
| **Genes** | **Forward** | **Reverse** |
| ***CTLA4*** | F; 5'-TCCCGCCCTACTACGGAAA-3' | R; 3'-GTAGGGATCCATGAAGCAGAGG-5' |
| ***Il4*** | F; 5'-TCTTTGCTGCCTCCAAGAACA-3' | R; 3'-TGTCGAGCCGTTTCAGGAAT-5' |
| ***Il5*** | F; 5'-GTGTATGCCATCCCCACAGA-3' | R; 3'-CTCTCCAGTGTGCCTATTCCC-5' |
| ***Il10*** | F; 5'-GACTTTAAGGGTTACCTGGGTTG-3' | R; 3'-TCTTGGTTCTCAGCTTGGGG-5' |
| ***GAPDH*** | F; 5'-GAGAAGGCTGGGGCTCATTT-3' | R; 3'-TAAGCAGTTGGTGGTGCAGG-5' |

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| **Table S2. Immunological findings at the time of diagnosis of immunodeficiency** | | | |
| **Immunologic data** | **Patients** | | |
| **LRBA (N=12)** | **CVID (N=12)** | ***p-value*** |
| **IgG, mg/dL** | 320 (111-548) | 113 (17-305) | 0.09 |
| **IgA, mg/dL** | 8.5 (1.7-43.7) | 7.0 (4.2-32.0) | 0.92 |
| **IgM, mg/dL** | 56.5 (25.7-151.0) | 17.0 (5.5-20.0) | <0.01\* |
| **IgE, IU/mL** | 0.6 (0.0-2.8) | 1.0 (0.7-12.5) | 0.25 |
| **WBC, cell/µL** | 7805 (5342-8697) | 7003 (5600-8210) | 0.64 |
| **Neutrophil, cell/µL** | 3922 (3136-6024) | 4200 (3587-5350) | 0.82 |
| **Lymphocyte, cell/µL** | 2235 (2035-2934) | 2150 (1800-4320) | 0.71 |
| **CD3+ T cells, cell/µL** | 1717 (1492-1765) | 1908 (1630-3214) | 0.36 |
| **CD4+ T cells, cell/µL** | 649 (451-882) | 833 (625-1917) | 0.12 |
| **CD8+ T cells, cell/µL** | 967 (670-1554) | 1175 (777-1406) | 0.62 |
| **CD19+ B cells, cell/µL** | 147 (93-451) | 111 (70-384) | 0.51 |
| *LRBA; LPS-Responsive-Beige-like Anchor, CVID; Common variable immune deficiency, N; Count, Ig; Immunoglobulin, WBC; White blood cell.*  *The median is shown [with 25th and 75th percentiles]. Mann-Whitney U test was used.*  *\* p-value is statistically significant <0.05* | | | |

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| **Table S3. History of clinical complications in patients with CVID and LRBA deficiency** | | | |
| **Type of complication** | **Patients** | | |
| **LRBA (N=12)** | **CVID (N=12)** | ***p-value*** |
| **Pneumonia (%)** | 9 (75.0) | 5 (41.7) | 0.09 |
| **Otitis media (%)** | 7 (58.3) | 5 (41.7) | 0.41 |
| **Sinusitis (%)** | 8 (66.7) | 7 (58.3) | 1.0 |
| **meningitis (%)** | 2 (16.7) | 0 (0.0) | 0.47 |
| **Skin infection (%)** | 1 (8.3) | 4 (33.3) | 0.31 |
| **Bronchiectasis (%)** | 7 (58.3) | 1 (8.3) | 0.02\* |
| **Osteomyelitis (%)** | 1 (8.3) | 0 (0.0) | 1.0 |
| **Septicemia (%)** | 1 (8.3) | 0 (0.0) | 1.0 |
| **Oral candidiasis (%)** | 3 (25.0) | 1 (8.3) | 0.59 |
| **Arthritis (%)** | 3 (25.0) | 2 (16.7) | 1.0 |
| **Chronic diarrhea (%)** | 10 (83.3) | 6 (50.0) | 0.19 |
| **Lymphadenopathy (%)** | 7 (58.3) | 6 (50.0) | 0.68 |
| **Splenomegaly (%)** | 9 (75.0) | 4 (33.3) | 0.04\* |
| **Hepatomegaly (%)** | 7 (58.3) | 2 (16.7) | 0.04\* |
| **Granulomas (%)** | 4 (33.3) | 0 (0.0) | 0.09 |
| **Failure to thrive (%)** | 4 (33.0) | 2 (16.7) | 0.64 |
| **Allergy (%)** | 2 (16.7) | 0 (0.0) | 0.4 |
| **Enteropathy (%)** | 9 (75.0) | 1 (8.3) | <0.01\* |
| **Autoimmunity (%)** | 8 (66.7) | 0 (0.0) | <0.01\* |
| *LRBA; LPS-Responsive-Beige-like Anchor, CVID; Common variable immune deficiency, N; Count.*  *The median is shown [with 25th and 75th percentiles]. Mann-Whitney U test was used.*  *\* p-value is statistically significant <0.05* | | | |

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| **Table S4. Changes in absolute count of lymphocyte and CD4+ T cell at time of immunodeficiency diagnosis with current study** | | | | | | |
| **Cells** | **Patients** | | | | | |
| **LRBA (N=12)** | | | **CVID (N=12)** | | |
| **Time of diagnosis** | **Current study** | ***p-value*** | **Time of diagnosis** | **Current study** | ***p-value*** |
| **Lymphocyte, cell/µL** | 2235 (2035-2934) | 2370 (1800-3400) | <0.01\* | 2150 (1800-4320) | 2050 (1375-2800) | <0.01\* |
| **CD4+ T cells, cell/µL** | 649 (451-882) | 694 (433-1022) | 0.92 | 833 (625-1917) | 585 (375-770) | 0.07 |
| *LRBA; LPS-Responsive-Beige-like Anchor, CVID; Common variable immune deficiency, N; Count,*  *The median is shown [with 25th and 75th percentiles]. Wilcoxon Signed Ranks Test was used.*  *\* p-value is statistically significant <0.05* | | | | | | |



**Figure S1. Flow cytometry analysis of CD4+ T cells.** Three examples demonstrate the ﬂow cytometric analysis of peripheral blood samples from HC, LRBA patient and CVID patient. PBMCs were isolated and stained with anti-CD4 PerCP-cy5.5. The cells were gated on lymphocytes for analysis of the CD4+ cells. Data are expressed as the median values of 13 participants in each group. Arrows indicate the gated population subsequently analyzed. LRBA; LPS responsive beige-like anchor protein, CVID; Common variable immunodeficiency, HC; Healthy controls.