

SUPPLEMENTARY MATERIAL

Table S1. Autoimmune manifestations between CVID patients With B lymphopenia and normal B cells.

Parameters	All CVID patients (n=387)	CVID with B lymphopenia (n=168)	CVID with normal B cells (n=219)	p-value
Autoimmunity (total)	94	53	41	0.003*
Hematologic autoimmunity	29	17	12	0.08
Immune thrombocytopenic purpura (ITP)	25	14	11	0.18
Autoimmune hemolytic anemia (AIHA)	15	10	5	0.06
Evans syndrome	11	7	4	0.20
Autoimmune neutropenia	1	1	0	0.44
Rheumatologic autoimmunity	30	17	13	0.12
Rheumatoid arthritis (RA)	13	8	4	0.13
Juvenile idiopathic arthritis (JIA)	14	8	6	0.41
Sjogren's syndrome	1	0	1	1.0
Systemic lupus erythematosus (SLE)	3	3	0	0.08
Autoimmune vasculitis	1	1	0	0.44
Kawasaki syndrome	2	1	1	1.0
Uveitis	3	1	2	1.0
Juvenile dermatomyositis (JDM)	1	1	0	0.44
Gastrointestinal autoimmunity	23	13	10	0.19
Inflammatory bowel disease (IBD)	9	5	4	0.51
Ulcerative colitis (UC)	4	2	2	1.0
Crohn's disease (CD)	2	0	2	0.50
Celiac disease	9	5	4	0.51
Autoimmune hepatitis	5	4	1	0.17
Dermatologic autoimmunity	18	7	11	0.69
Alopecia areata	6	3	3	1.0
Vitiligo	8	4	4	0.73
Lichen planus (LP)	2	0	2	0.50
Psoriasis	3	1	2	1.0
Endocrine autoimmunity	6	4	2	0.24
Autoimmune thyroid disease	3	2	1	0.58
Hashimoto's Thyroiditis	2	1	1	1.0
Growth hormone deficiency	1	1	0	0.44
Neurological autoimmunity	5	1	4	0.28
Guillain-Barre syndrome	3	1	2	1.0
Myasthenia gravis (MG)	2	0	2	0.5

n; Number * *p*-value is statistically significant <0.05

Table S2. Lymphoproliferative diseases by locations between CVID patients With B lymphopenia and normal B cells.

Parameters	All CVID patients (n=387)	CVID with B lymphopenia (n=168)	CVID with normal B cells (n=219)	<i>p</i> -value
Lymphoproliferation	83 (21.4%)	39 (23.2%)	44 (20.1%)	0.45
Splenomegaly	101 (26.1%)	62 (36.9%)	39 (17.8%)	<0.001*
Hepatomegaly	67 (17.3%)	43 (25.6%)	24 (11%)	<0.001*
Granulomas	8 (2.1%)	5 (3%)	3 (1.4%)	0.30
Liver	2	2	2	1.0
Skin	3	1	-	0.43
Brain	1	-	1	1.0
Lung	2	2	-	0.19
Lymph nodes	1	1	-	0.43
Spleen	1	1	-	0.43
<i>n</i> : Number				
* <i>p</i> -value is statistically significant <0.05				

Table S3. Other clinical manifestations comparison between CVID patients With B lymphopenia and normal B cells.

Parameters	All CVID patients (n=387)	CVID with B lymphopenia (n=168)	CVID with normal B cells (n=219)	p-value
Hematological manifestations (%)	72 (18.6%)	42 (25%)	30 (13.7%)	0.004*
Pancytopenia	19 (4.9%)	11 (6.5%)	8 (3.6%)	0.19
Anemia	49 (12.6%)	23 (13.6%)	26 (11.8%)	0.59
Neutropenia	30 (7.7%)	17 (10.11%)	13 (5.9%)	0.99
Thrombocytopenia	38 (9.8%)	20 (11.9%)	18 (8.2%)	0.22
Leukopenia	13 (3.3%)	5 (2.9%)	8 (3.6%)	0.78
Other clinical manifestations				
Failure to thrive (%)	76 (19.6%)	37 (22%)	39 (17.8%)	0.30
Bronchiectasis (%)	99 (25.6%)	57 (33.9%)	42 (19.2%)	<0.001*
Clubbing (%)	70 (18.1%)	41 (24.4%)	29 (13.2%)	0.004*
Oral ulcer (%)	49 (12.7%)	21 (12.5%)	28 (12.8%)	0.93
Sterile meningitis (%)	11 (2.8%)	5 (3%)	6 (2.7%)	1.0
Sterile abscess (%)	47 (12.1%)	21 (12.5%)	26 (11.9%)	0.85
Sterile conjunctivitis (%)	42 (10.9%)	25 (14.9%)	17 (7.8%)	0.02*
<i>n</i> ; Number				
* <i>p</i> -value is statistically significant <0.05				

Table S4. Malignancy types between CVID patients With B lymphopenia and normal B cells.

Parameters	All CVID patients (n=387)	CVID with B lymphopenia (n=168)	CVID with normal B cells (n=219)	<i>p</i> -value
Malignancy (total)	28 (7.2%)	14 (8.3%)	13 (5.9%)	0.35
Hematologic cancers	24 (6.2%)	12 (7.1%)	12 (5.4%)	0.50
Lymphoma (total)	22 (5.6%)	11 (6%)	11 (5%)	0.52
Hodgkin lymphoma	8 (2%)	4 (2%)	4 (1.8%)	0.73
Non- Hodgkin lymphoma	14 (3.6%)	7 (4.1%)	7 (3.1%)	0.61
Leukemia	2 (0.5%)	1 (0.5%)	1 (0.4%)	1.0
Solid tumors	4 (1%)	3 (1.7%)	1 (0.4%)	0.20
Breast cancer	1 (0.2%)	-	1 (0.4%)	1.0
Ovarian cancer	1 (0.2%)	1 (0.5%)	-	0.43
Brain cancer	1 (0.2%)	1 (0.5%)	-	0.43
Gastric cancer	1 (0.2%)	1 (0.5%)	-	0.43
<i>n</i> : Number				
* <i>p</i> -value is statistically significant <0.05				

Table S5. PFT parameters and CT scan analysis between CVID patients With B lymphopenia and normal B cells.

PFT results	All CVID patients (n=65)	CVID with B lymphopenia (n=32)	CVID with normal B cells (n=33)	p-value
Normal	30 (46.2%)	11 (34.3%)	19 (57.5%)	0.47
Obstructive	12 (18.5%)	7 (21.8%)	5 (15.1%)	0.17
Restrictive	14 (21.5%)	8 (25.0%)	6 (18.1%)	0.15
Mixed	9 (13.8%)	6 (18.7%)	3 (9.1%)	0.08
PFT value, % (IQR)				
PEF	64.2 (46.1-81.5)	61.8 (47-84.5)	64.4 (50.2-80.0)	0.53
FVC	85.7 (67.7-102.7)	82.0 (62.0-104.2)	79.2 (71.5-100.5)	0.12
FEV1	79.6 (55.5-100.4)	71.6 (50.5-95.9)	76.1 (67.7-101.0)	0.48
MEF25-75	63.3 (31.4-85.2)	51.7 (29.5-72.1)	69.8 (55.5-86.5)	0.09
HRCT results	All CVID patients (n=70)	CVID with B lymphopenia (n=32)	CVID with normal B cells (n=38)	p-value
Normal	18 (25.7%)	6 (18.7%)	12 (31.5%)	0.22
Abnormal	52 (74.2%)	26 (81.2%)	26 (68.4%)	
<i>n; Number, Abbreviations: PFT, Pulmonary function testing; HRCT, high-resolution computed tomography; PEF, peak expiratory flow; FEV1, forced expiratory volume; FVC, forced volume vital capacity; MEF, maximal expiratory flow. * p-value is statistically significant <0.05</i>				