Stephan Ehl

List of Publications by Year
in descending order

202	13,577	22132	25770
papers	citations	59 h-index	108 g-index

206	206	206	14942
all docs	docs citations	times ranked	citing authors

#	Article	IF	Citations
1	Autosomal dominant immune dysregulation syndrome in humans with CTLA4 mutations. Nature Medicine, 2014, 20, 1410-1416.	15.2	723
2	Antigen localisation regulates immune responses in a dose- and time-dependent fashion: a geographical view of immune reactivity. Immunological Reviews, 1997, 156, 199-209.	2.8	469
3	The European Society for Immunodeficiencies (ESID) Registry Working Definitions for the ClinicalÂDiagnosis of Inborn Errors of Immunity. Journal of Allergy and Clinical Immunology: in Practice, 2019, 7, 1763-1770.	2.0	381
4	Clinical spectrum and features of activated phosphoinositide 3-kinase Î´ syndrome: AÂlarge patient cohort study. Journal of Allergy and Clinical Immunology, 2017, 139, 597-606.e4.	1.5	377
5	Selective predisposition to bacterial infections in IRAK-4â€“deficient children: IRAK-4â€“dependent TLRs are otherwise redundant in protective immunity. Journal of Experimental Medicine, 2007, 204, 2407-2422.	4.2	374
6	Familial Hemophagocytic Lymphohistiocytosis Type 5 (FHL-5) Is Caused by Mutations in Munc18-2 and Impaired Binding to Syntaxin 11. American Journal of Human Genetics, 2009, 85, 482-492.	2.6	370
7	Lambda Interferon Renders Epithelial Cells of the Respiratory and Gastrointestinal Tracts Resistant to Viral Infections. Journal of Virology, 2010, 84, 5670-5677.	1.5	369
8	Clinical Features and Outcome of Patients With IRAK-4 and MyD88 Deficiency. Medicine (United States), 2010, 89, 403-425.	0.4	366
9	Phenotype, penetrance, and treatment of 133 cytotoxic T-lymphocyte antigen 4â€“insufficient subjects. Journal of Allergy and Clinical Immunology, 2018, 142, 1932-1946.	1.5	344
10	ORAI1 deficiency and lack of store-operated Ca2+ entry cause immunodeficiency, myopathy, and ectodermal dysplasia. Journal of Allergy and Clinical Immunology, 2009, 124, 1311-1318.e7.	1.5	289
11	Interleukin-18 diagnostically distinguishes and pathogenically promotes human and murine macrophage activation syndrome. Blood, 2018, 131, 1442-1455.	0.6	288
12	DOCK8 Deficiency: Clinical and Immunological Phenotype and Treatment Options - a Review of 136 Patients. Journal of Clinical Immunology, 2015, 35, 189-198.	2.0	284
13	A prospective evaluation of degranulation assays in the rapid diagnosis of familial hemophagocytic syndromes. Blood, 2012, 119, 2754-2763.	0.6	263
14	Long-term follow-up of IPEX syndrome patients after different therapeutic strategies: An international multicenter retrospective study. Journal of Allergy and Clinical Immunology, 2018, 141, 1036-1049.e5.	1.5	233
15	Lethal hemophagocytic lymphohistiocytosis in Hermansky-Pudlak syndrome type II. Blood, 2006, 108, 81-87.	0.6	194
16	ORAI1-mediated calcium influx is required for human cytotoxic lymphocyte degranulation and target cell lysis. Proceedings of the National Academy of Sciences of the United States of America, 2011, 108, 3324-3329.	3.3	181
17	Dendritic Cells Efficiently Induce Protective Antiviral Immunity. Journal of Virology, 1998, 72, 3812-3818.	1.5	175
18	Oncogenic JAK2 ^{V617F} causes PD-L1 expression, mediating immune escape in myeloproliferative neoplasms. Science Translational Medicine, 2018, 10, .	5.8	166

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19	Bystander Activation of Cytotoxic T Cells: Studies on the Mechanism and Evaluation of In Vivo Significance in a Transgenic Mouse Model. Journal of Experimental Medicine, 1997, 185, 1241-1252.	4.2	165
20	Deficiency of Innate and Acquired Immunity Caused by an IKBKB Mutation. New England Journal of Medicine, 2013, 369, 2504-2514.	13.9	161
21	The syndrome of hemophagocytic lymphohistiocytosis in primary immunodeficiencies: implications for differential diagnosis and pathogenesis. Haematologica, 2015, 100, 978-988.	1.7	161
22	Viral and Bacterial Infections Interfere with Peripheral Tolerance Induction and Activate CD8+ T Cells to Cause Immunopathology. Journal of Experimental Medicine, 1998, 187, 763-774.	4.2	158
23	Clinical and immunological manifestations of patients with atypical severe combined immunodeficiency. Clinical Immunology, 2011, 141, 73-82.	1.4	157
24	Antiviral and Regulatory T Cell Immunity in a Patient with Stromal Interaction Molecule 1 Deficiency. Journal of Immunology, 2012, 188, 1523-1533.	0.4	156
25	PGM3 Mutations Cause a Congenital Disorder of Glycosylation with Severe Immunodeficiency and Skeletal Dysplasia. American Journal of Human Genetics, 2014, 95, 96-107.	2.6	148
26	Mutations in AP3D1 associated with immunodeficiency and seizures define a new type of Hermansky-Pudlak syndrome. Blood, 2016, 127, 997-1006.	0.6	142
27	A variant of SCID with specific immune responses and predominance of ÂÂ T cells. Journal of Clinical Investigation, 2005, 115, 3140-3148.	3.9	139
28	Distinct mutations in STXBP2 are associated with variable clinical presentations in patients with familial hemophagocytic lymphohistiocytosis type 5 (FHL5). Blood, 2012, 119, 6016-6024.	0.6	137
29	Diseaseâ€causing mutations in the <scp>XIAP</scp> <scp>BIR</scp> 2 domain impair <scp>NOD</scp> 2â€dependent immune signalling. EMBO Molecular Medicine, 2013, 5, 1278-1295.	3.3	137
30	Disease Evolution and Response to Rapamycin in Activated Phosphoinositide 3-Kinase Î´ Syndrome: The European Society for Immunodeficiencies-Activated Phosphoinositide 3-Kinase Î´ Syndrome Registry. Frontiers in Immunology, 2018, 9, 543.	2.2	137
31	Recent advances in the diagnosis and treatment of hemophagocytic lymphohistiocytosis. Arthritis Research and Therapy, 2012, 14, 213.	1.6	129
32	A Severe Form of Human Combined Immunodeficiency Due to Mutations in DNA Ligase IV. Journal of Immunology, 2006, 176, 5060-5068.	0.4	128
33	Malignancyâ€associated haemophagocytic lymphohistiocytosis in children and adolescents. British Journal of Haematology, 2015, 170, 539-549.	1.2	118
34	Hypomorphic caspase activation and recruitment domain 11 (CARD11) mutations associated with diverse immunologic phenotypes with or without atopic disease. Journal of Allergy and Clinical Immunology, 2019, 143, 1482-1495.	1.5	116
35	Virus clearance and immunopathology by CD8+ T cells during infection with respiratory syncytial virus are mediated by IFN-Î³. European Journal of Immunology, 2002, 32, 2117.	1.6	113
36	Recommendations for the Use of Etoposide-Based Therapy and Bone Marrow Transplantation for the Treatment of HLH: Consensus Statements by the HLH Steering Committee of the Histiocyte Society. Journal of Allergy and Clinical Immunology: in Practice, 2018, 6, 1508-1517.	2.0	112

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37	Atypical familial hemophagocytic lymphohistiocytosis due to mutations in UNC13D and STXBP2 overlaps with primary immunodeficiency diseases. Haematologica, 2010, 95, 2080-2087.	1.7	109
38	Diagnostic evaluation of patients with suspected haemophagocytic lymphohistiocytosis. British Journal of Haematology, 2013, 160, 275-287.	1.2	109
39	Hematopoietic stem cell transplantation in 29 patients hemizygous for hypomorphic IKBKG/NEMO mutations. Blood, 2017, 130, 1456-1467.	0.6	95
40	The role of Toll-like receptor 4 versus interleukin-12 in immunity to respiratory syncytial virus. European Journal of Immunology, 2004, 34, 1146-1153.	1.6	93
41	Hematopoietic stem cell transplant in patients with activated PI3K delta syndrome. Journal of Allergy and Clinical Immunology, 2017, 139, 1046-1049.	1.5	90
42	A functional and kinetic comparison of antiviral effector and memory cytotoxic T lymphocyte populationsin vivo andin vitro. European Journal of Immunology, 1997, 27, 3404-3413.	1.6	85
43	Hemophagocytic lymphohistiocytosis in syntaxin-11â€“deficient mice: T-cell exhaustion limits fatal disease. Blood, 2013, 121, 604-613.	0.6	85
44	Long-term persistence and reactivation of T cell memory in the lung of mice infected with respiratory syncytial virus. European Journal of Immunology, 2001, 31, 2574-2582.	1.6	84
45	The minimum required level of donor chimerism in hereditary hemophagocytic lymphohistiocytosis. Blood, 2016, 127, 3281-3290.	0.6	83
46	Hemophagocytic lymphohistiocytosis in adults: collaborative analysis of 137 cases of a nationwide German registry. Journal of Cancer Research and Clinical Oncology, 2020, 146, 1065-1077.	1.2	83
47	The expansion of human T-bet ^{high} CD21 ^{low} B cells is T cell dependent. Science Immunology, 2021, 6, eabh0891.	5.6	82
48	Direct quantitation of rapid elimination of viral antigen-positive lymphocytes by antiviral CD8+ T cellsin vivo. European Journal of Immunology, 2000, 30, 1356-1363.	1.6	78
49	Subtle differences in CTL cytotoxicity determine susceptibility to hemophagocytic lymphohistiocytosis in mice and humans with Chediak-Higashi syndrome. Blood, 2011, 118, 4620-4629.	0.6	78
50	Hyperactive mTOR pathway promotes lymphoproliferation and abnormal differentiation in autoimmune lymphoproliferative syndrome. Blood, 2016, 128, 227-238.	0.6	77
51	Initial presenting manifestations in 16,486 patients with inborn errors of immunity include infections and noninfectious manifestations. Journal of Allergy and Clinical Immunology, 2021, 148, 1332-1341.e5.	1.5	75
52	Early-onset autoimmunity associated with SOCS1 haploinsufficiency. Nature Communications, 2020, 11, 5341.	5.8	74
53	The risk of hemophagocytic lymphohistiocytosis in Hermansky-Pudlak syndrome type 2. Blood, 2013, 121, 2943-2951.	0.6	72
54	A prospective study on the natural history of patients with profound combined immunodeficiency: An interim analysis. Journal of Allergy and Clinical Immunology, 2017, 139, 1302-1310.e4.	1.5	71

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55	Syntaxin binding mechanism and disease-causing mutations in Munc18-2. Proceedings of the National Academy of Sciences of the United States of America, 2013, 110, E4482-91.	3.3	70
56	Primary and secondary hemophagocytic lymphohistiocytosis have different patterns of Tâ€cell activation, differentiation and repertoire. European Journal of Immunology, 2017, 47, 364-373.	1.6	69
57	Early-onset Evans syndrome, immunodeficiency, and premature immunosenescence associated with tripeptidyl-peptidase II deficiency. Blood, 2015, 125, 753-761.	0.6	66
58	Î²2-Microglobulin deficiency causes a complex immunodeficiency of the innate and adaptive immune system. Journal of Allergy and Clinical Immunology, 2015, 136, 392-401.	1.5	66
59	A comparison of efficacy and specificity of three NK depleting antibodies. Journal of Immunological Methods, 1996, 199, 149-153.	0.6	63
60	Reduced memory B cells in patients with hyper IgE syndrome. Clinical Immunology, 2008, 129, 448-454.	1.4	63
61	Two siblings with lethal pneumococcal meningitis in a family with a mutation in Interleukin-1 receptorâ€“associated kinase 4. Journal of Pediatrics, 2004, 145, 698-700.	0.9	62
62	ORAI1 mutations abolishing store-operated Ca2+ entry cause anhidrotic ectodermal dysplasia with immunodeficiency. Journal of Allergy and Clinical Immunology, 2018, 142, 1297-1310.e11.	1.5	62
63	Clinical and immunologic consequences of a somatic reversion in a patient with X-linked severe combined immunodeficiency. Blood, 2008, 112, 4090-4097.	0.6	59
64	Graded Defects in Cytotoxicity Determine Severity of Hemophagocytic Lymphohistiocytosis in Humans and Mice. Frontiers in Immunology, 2013, 4, 448.	2.2	59
65	Germline TET2 loss of function causes childhood immunodeficiency and lymphoma. Blood, 2020, 136, 1055-1066.	0.6	58
66	Antigen persistence and time of T-cell tolerization determine the efficacy of tolerization protocols for prevention of skin graft rejection. Nature Medicine, 1998, 4, 1015-1019.	15.2	56
67	Tyrosine kinase 2 is not limiting human antiviral type III interferon responses. European Journal of Immunology, 2016, 46, 2639-2649.	1.6	56
68	The most frequentDCLRE1C(ARTEMIS) mutations are based on homologous recombination events. Human Mutation, 2010, 31, 197-207.	1.1	55
69	Numerical modelling of label-structured cell population growth using CFSE distribution data. Theoretical Biology and Medical Modelling, 2007, 4, 26.	2.1	54
70	Abnormally differentiated CD4+ or CD8+ T cells with phenotypic and genetic features of double negative T cells in human Fas deficiency. Blood, 2014, 124, 851-860.	0.6	54
71	Quorum Regulation via Nested Antagonistic Feedback Circuits Mediated by the Receptors CD28 and CTLA-4 Confers Robustness to T Cell Population Dynamics. Immunity, 2020, 52, 313-327.e7.	6.6	54
72	Missense mutation in immunodeficient patients shows the multifunctional roles of coiled-coil domain 3 (CC3) in STIM1 activation. Proceedings of the National Academy of Sciences of the United States of America, 2015, 112, 6206-6211.	3.3	52

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73	Treatment of severe forms of LPS-responsive beige-like anchor protein deficiency with allogeneic hematopoietic stem cell transplantation. Journal of Allergy and Clinical Immunology, 2018, 141, 770-775.e1.	1.5	52
74	A cautionary note on experimental artefacts induced by fetal calf serum in a viral model of pulmonary eosinophilia. Journal of Immunological Methods, 2002, 268, 211-218.	0.6	51
75	General and specific immunosuppression caused by antiviral T-cell responses. Immunological Reviews, 1999, 168, 305-315.	2.8	49
76	Clinical, radiographic, and genetic diagnosis of progressive pseudorheumatoid dysplasia in a patient with severe polyarthropathy. Rheumatology International, 2004, 24, 53-56.	1.5	49
77	Functional impairment of cytotoxic T cells in the lung airways following respiratory virus infections. European Journal of Immunology, 2006, 36, 1434-1442.	1.6	49
78	Evolution of disease activity and biomarkers on and off rapamycin in 28 patients with autoimmune lymphoproliferative syndrome. Haematologica, 2017, 102, e52-e56.	1.7	49
79	Exhaustion of cytotoxic T cells during adoptive immunotherapy of virus carrier mice can be prevented by B cells or CD4+ T cells. European Journal of Immunology, 2002, 32, 374-382.	1.6	48
80	NCKAP1L defects lead to a novel syndrome combining immunodeficiency, lymphoproliferation, and hyperinflammation. Journal of Experimental Medicine, 2020, 217, .	4.2	48
81	Human metapneumovirus induces more severe disease and stronger innate immune response in BALB/c mice as compared with respiratory syncytial virus. Respiratory Research, 2007, 8, 6.	1.4	47
82	The Impact of Variation in the Number of CD8+T-Cell Precursors on the Outcome of Virus Infection. Cellular Immunology, 1998, 189, 67-73.	1.4	46
83	Hemophagocytic Lymphohistiocytosis in Imported Pediatric Visceral Leishmaniasis in a Nonendemic Area. Journal of Pediatrics, 2014, 165, 147-153.e1.	0.9	46
84	Delayed-onset adenosine deaminase deficiency: Strategies for an early diagnosis. Journal of Allergy and Clinical Immunology, 2012, 130, 991-994.	1.5	44
85	T-cell gene therapy for perforin deficiency correctsÂcytotoxicity defects and prevents hemophagocytic lymphohistiocytosis manifestations. Journal of Allergy and Clinical Immunology, 2018, 142, 904-913.e3.	1.5	44
86	Severe eczema and Hyper-IgE in Loeysâ€“Dietz-syndrome â€" Contribution to new findings of immune dysregulation in connective tissue disorders. Clinical Immunology, 2014, 150, 43-50.	1.4	43
87	Chronic Inflammatory Bowel Disease as Key Manifestation of Atypical ARTEMIS Deficiency. Journal of Clinical Immunology, 2010, 30, 314-320.	2.0	42
88	Wiskottâ€“Aldrich syndrome presenting with a clinical picture mimicking juvenile myelomonocytic leukaemia. Pediatric Blood and Cancer, 2013, 60, 836-841.	0.8	42
89	Therapeutic strategy in p47-phox deficient chronic granulomatous disease presenting as inflammatory bowel disease. Journal of Allergy and Clinical Immunology, 2010, 125, 943-946.e1.	1.5	40
90	Prevalence and clinical challenges among adults with primary immunodeficiency and recombination-activating gene deficiency. Journal of Allergy and Clinical Immunology, 2018, 141, 2303-2306.	1.5	40

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91	Distinct molecular response patterns of activating STAT3 mutations associate with penetrance of lymphoproliferation and autoimmunity. Clinical Immunology, 2020, 210, 108316.	1.4	40
92	Multisystem inflammation and susceptibility to viral infections in human ZNFX1 deficiency. Journal of Allergy and Clinical Immunology, 2021, 148, 381-393.	1.5	40
93	Platelet secretion defect in patients with familial hemophagocytic lymphohistiocytosis type 5 (FHL-5). Blood, 2010, 116, 6148-6150.	0.6	39
94	Patients with T^{+/low} NK⁺ IL2 receptor Î³ chain deficiency have differentiallyâ€impaired cytokine signaling resulting in severe combined immunodeficiency. European Journal of Immunology, 2014, 44, 3129-3140.	1.6	39
95	XIAP deficiency is a mendelian cause of late-onset IBD. Gut, 2014, 63, 1031-1032.	6.1	38
96	SYK expression endows human ZAP70-deficient CD8 T cells with residual TCR signaling. Clinical Immunology, 2015, 161, 103-109.	1.4	38
97	Transplantation from a symptomatic carrier sister restores host defenses but does not prevent colitis in NEMO deficiency. Clinical Immunology, 2016, 164, 52-56.	1.4	38
98	"Bystander" recruitment of systemic memory Tâ€„cells delays the immune response to respiratory virus infection. European Journal of Immunology, 2003, 33, 1839-1848.	1.6	37
99	Computational analysis of CFSE proliferation assay. Journal of Mathematical Biology, 2006, 54, 57-89.	0.8	37
100	Common variable immunodeficiency in children. Current Opinion in Pediatrics, 2007, 19, 685-692.	1.0	37
101	Omenn syndrome associated with a functional reversion due to a somatic second-site mutation in CARD11 deficiency. Blood, 2015, 126, 1658-1669.	0.6	37
102	Effective Immunological Guidance of Genetic Analyses Including Exome Sequencing in Patients Evaluated for Hemophagocytic Lymphohistiocytosis. Journal of Clinical Immunology, 2017, 37, 770-780.	2.0	37
103	Rapid identification and characterization of infected cells in blood during chronic active Epstein-Barr virus infection. Journal of Experimental Medicine, 2020, 217, .	4.2	37
104	Role of T Cells in Virus Control and Disease after Infection with Pneumonia Virus of Mice. Journal of Virology, 2008, 82, 11619-11627.	1.5	36
105	Lesson from hypomorphic recombination-activating gene (RAG) mutations: Why asymptomatic siblings should also be tested. Journal of Allergy and Clinical Immunology, 2014, 133, 1211-1215.e2.	1.5	36
106	Evaluating laboratory criteria for combined immunodeficiency in adult patients diagnosed with common variable immunodeficiency. Clinical Immunology, 2019, 203, 59-62.	1.4	36
107	International retrospective study of allogeneic hematopoietic cell transplantation for activated PI3K-delta syndrome. Journal of Allergy and Clinical Immunology, 2022, 149, 410-421.e7.	1.5	34
108	Clinical Heterogeneity of Immunodysregulation, Polyendocrinopathy, Enteropathy, X-linked: Pulmonary Involvement as a Non-Classical Disease Manifestation. Journal of Clinical Immunology, 2014, 34, 601-606.	2.0	33

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109	Neuroinflammatory Disease as an Isolated Manifestation of Hemophagocytic Lymphohistiocytosis. Journal of Clinical Immunology, 2020, 40, 901-916.	2.0	33
110	Low level viral persistence after infection with LCMV: a quantitative insight through numerical bifurcation analysis. Mathematical Biosciences, 2001, 173, 1-23.	0.9	30
111	A novel single point mutation of the LYST gene in two siblings with different phenotypic features of Chediak Higashi syndrome. Pediatric Blood and Cancer, 2011, 56, 1136-1139.	0.8	30
112	Targeted busulfan-based reduced-intensity conditioning and HLA-matched HSCT cure hemophagocytic lymphohistiocytosis. Blood Advances, 2020, 4, 1998-2010.	2.5	30
113	Sequential decisions on FAS sequencing guided by biomarkers in patients with lymphoproliferation and autoimmune cytopenia. Haematologica, 2013, 98, 1948-1955.	1.7	29
114	<scp>CD</scp>57 identifies T cells with functional senescence before terminal differentiation and relative telomere shortening in patients with activated <scp>PI</scp>3 kinase delta syndrome. Immunology and Cell Biology, 2018, 96, 1060-1071.	1.0	29
115	Retroviral UNC13D Gene Transfer Restores Cytotoxic Activity of T Cells Derived from Familial Hemophagocytic Lymphohistiocytosis Type 3 Patients In Vitro. Human Gene Therapy, 2019, 30, 975-984.	1.4	29
116	Pulmonary T cells induced by respiratory syncytial virus are functional and can make an important contribution to long-lived protective immunity. European Journal of Immunology, 2002, 32, 2562-2569.	1.6	27
117	Differences in Granule Morphology yet Equally Impaired Exocytosis among Cytotoxic T Cells and NK Cells from Chediakâ€“Higashi Syndrome Patients. Frontiers in Immunology, 2017, 8, 426.	2.2	26
118	Is an infectious trigger always required for primary hemophagocytic lymphohistiocytosis? Lessons from in utero and neonatal disease. Pediatric Blood and Cancer, 2018, 65, e27344.	0.8	26
119	Disturbed B-lymphocyte selection in autoimmune lymphoproliferative syndrome. Blood, 2016, 127, 2193-2202.	0.6	25
120	Germline STAT3 gain-of-function mutations in primary immunodeficiency: Impact on the cellular and clinical phenotype. Biomedical Journal, 2021, 44, 412-421.	1.4	25
121	A distinct CD38+CD45RA+ population of CD4+, CD8+, and double-negative T cells is controlled by FAS. Journal of Experimental Medicine, 2021, 218, .	4.2	25
122	Recombinant Sendai virus induces T cell immunity against respiratory syncytial virus that is protective in the absence of antibodies. Cellular Immunology, 2007, 247, 85-94.	1.4	24
123	Hemophagocytic lymphohistiocytosis as presenting manifestation of profound combined immunodeficiency due to an ORAI1 mutation. Journal of Allergy and Clinical Immunology, 2017, 140, 1721-1724.	1.5	23
124	Cell Versus Cytokine â€“ Directed Therapies for Hemophagocytic Lymphohistiocytosis (HLH) in Inborn Errors of Immunity. Frontiers in Immunology, 2020, 11, 808.	2.2	23
125	Both Nonstructural Proteins NS1 and NS2 of Pneumonia Virus of Mice are Inhibitors of the Interferon Type I and Type III Responses In Vivo. Journal of Virology, 2011, 85, 4071-4084.	1.5	22
126	A RAB27A 5â€² untranslated region structural variant associated with late-onset hemophagocytic lymphohistiocytosis and normal pigmentation. Journal of Allergy and Clinical Immunology, 2018, 142, 317-321.e8.	1.5	22

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127	Gray platelet syndrome can mimic autoimmune lymphoproliferative syndrome. Blood, 2015, 126, 1967-1969.	0.6	21
128	Seletalisib for Activated PI3KÎ´ Syndromes: Open-Label Phase 1b and Extension Studies. Journal of Immunology, 2020, 205, 2979-2987.	0.4	21
129	Modelling the Dynamics of LCMV Infection in Mice: II. Compartmental Structure and Immunopathology. Journal of Theoretical Biology, 2003, 221, 349-378.	0.8	20
130	Translating the Genomic Revolution â€" Targeted Genome Editing in Primates. New England Journal of Medicine, 2014, 370, 2342-2345.	13.9	20
131	Profound immunodeficiency with severe skin disease explained by concomitant novel CARMIL2 and PLEC1 loss-of-function mutations. Clinical Immunology, 2019, 208, 108228.	1.4	20
132	Donor cell persistence and activation-induced unresponsiveness of peripheral CD8+ T cells. European Journal of Immunology, 2000, 30, 883-891.	1.6	19
133	Establishing the Molecular Diagnoses in a Cohort of 291 Patients With Predominantly Antibody Deficiency by Targeted Next-Generation Sequencing: Experience From a Monocentric Study. Frontiers in Immunology, 2021, 12, 786516.	2.2	19
134	Novel mutation in Hermanskyâ€"Pudlak syndrome type 2 with mild immunological phenotype. Platelets, 2013, 24, 538-543.	1.1	18
135	Erythropoiesis defect observed in STAT3 GOF patients with severe anemia. Journal of Allergy and Clinical Immunology, 2020, 145, 1297-1301.	1.5	18
136	An improved protocol for measuring cytotoxic T cell activity in anatomic compartments with low cell numbers. Journal of Immunological Methods, 2001, 257, 155-161.	0.6	17
137	Major Histocompatibility Complex-Dependent Cytotoxic T Lymphocyte Repertoire and Functional Avidity Contribute to Strain-Specific Disease Susceptibility after Murine Respiratory Syncytial Virus Infection. Journal of Virology, 2011, 85, 10135-10143.	1.5	17
138	TIMâ€3 deficiency presenting with two clonally unrelated episodes of mesenteric and subcutaneous panniculitisâ€like Tâ€cell lymphoma and hemophagocytic lymphohistiocytosis. Pediatric Blood and Cancer, 2020, 67, e28302.	0.8	17
139	Preserved effector functions of human ORAI1- and STIM1-deficient neutrophils. Journal of Allergy and Clinical Immunology, 2016, 137, 1587-1591.e7.	1.5	16
140	T+ NK+ IL-2 Receptor Î³ Chain Mutation: a Challenging Diagnosis of Atypical Severe Combined Immunodeficiency. Journal of Clinical Immunology, 2018, 38, 527-536.	2.0	16
141	Influence of a Single Viral Epitope on T Cell Response and Disease After Infection of Mice with Respiratory Syncytial Virus. Journal of Immunology, 2007, 179, 8264-8273.	0.4	15
142	Reversible pancytopenia and immunodeficiency in a patient with hereditary folate malabsorption. Pediatric Blood and Cancer, 2015, 62, 1091-1094.	0.8	15
143	High Levels of IL-18 and IFN-Î³ in Chronically Inflamed Tissue in Chronic Granulomatous Disease. Frontiers in Immunology, 2019, 10, 2236.	2.2	15
144	Toll-Like Receptor Stimulation Induces Higher TNF-α Secretion in Peripheral Blood Mononuclear Cells from Patients with Hyper IgE Syndrome. International Archives of Allergy and Immunology, 2008, 146, 190-194.	0.9	14

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145	The NEW ESID online database network. Bioinformatics, 2019, 35, 5367-5369.	1.8	14
146	Griscelli Syndrome Type 2 Sine Albinism: Unraveling Differential RAB27A Effector Engagement. Frontiers in Immunology, 2020, 11, 612977.	2.2	14
147	Definition and validation of serum biomarkers for optimal differentiation of hyperferritinaemic cytokine storm conditions in children: a retrospective cohort study. Lancet Rheumatology, The, 2021, 3, e563-e573.	2.2	14
148	Agammaglobulinemia with normal B-cell numbers in a patient lacking Bob1. Journal of Allergy and Clinical Immunology, 2021, 147, 1977-1980.	1.5	12
149	Platelet secretion defect in a patient with stromal interaction molecule 1 deficiency. Blood, 2013, 122, 3696-3698.	0.6	11
150	Novel Patient with Late-Onset Familial Hemophagocytic Lymphohistiocytosis with STXBP2 Mutations Presenting with Autoimmune Hepatitis, Neurological Manifestations and Infections Associated with Hypogammaglobulinemia. Journal of Clinical Immunology, 2015, 35, 22-25.	2.0	11
151	The autoimmune targets in IPEX are dominated by gut epithelial proteins. Journal of Allergy and Clinical Immunology, 2019, 144, 327-330.e8.	1.5	11
152	Triggerâ€dependent differences determine therapeutic outcome in murine primary hemophagocytic lymphohistiocytosis. European Journal of Immunology, 2020, 50, 1770-1782.	1.6	11
153	Rubella vaccineâ€“induced granulomas are a novel phenotype with incomplete penetrance of genetic defects in cytotoxicity. Journal of Allergy and Clinical Immunology, 2022, 149, 388-399.e4.	1.5	11
154	Altered T-Lymphocyte Biology Following High-Dose Melphalan and Autologous Stem Cell Transplantation With Implications for Adoptive T-Cell Therapy. Frontiers in Oncology, 2020, 10, 568056.	1.3	11
155	CD59 deficiency presenting as polyneuropathy and Moyamoya syndrome with endothelial abnormalities of small brain vessels. European Journal of Paediatric Neurology, 2018, 22, 870-877.	0.7	10
156	Earlyâ€onset, fatal interstitial lung disease in STAT3 gainâ€ofâ€function patients. Pediatric Pulmonology, 2021, 56, 3934-3941.	1.0	9
157	Simple Measurement of IgA Predicts Immunity and Mortality in Ataxia-Telangiectasia. Journal of Clinical Immunology, 2021, 41, 1878-1892.	2.0	9
158	Reduced-Intensity/Reduced-Toxicity Conditioning Approaches Are Tolerated in XIAP Deficiency but Patients Fare Poorly with Acute GVHD. Journal of Clinical Immunology, 2021, , 1.	2.0	9
159	The impact of splenectomy on antiviral T cell memory in mice. International Immunology, 2004, 17, 27-33.	1.8	8
160	Massive monoclonal expansion of CD4 T-cells specific for aMycobacterium tuberculosisESAT-6 peptide. European Respiratory Journal, 2012, 40, 152-160.	3.1	8
161	Risk factors for mixed chimerism in children with hemophagocytic lymphohistiocytosis after reduced toxicity conditioning. Pediatric Blood and Cancer, 2020, 67, e28523.	0.8	8
162	Primary haemophagocytic lymphohistiocytosis (ChÃ©diakâ€Higashi Syndrome) triggered by acute SARSâ€CoVâ€2 infection in a sixâ€weekâ€old infant. British Journal of Haematology, 2021, 195, 198-200.	1.2	8

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