V Koneti Rao

List of Publications by Year
in descending order

90 papers	6,618 citations	109321 35 h-index	64796 79 g-index
94 all docs	94 docs citations	94 times ranked	8051 citing authors

#	Article	IF	Citations
1	Immune dysregulation in human subjects with heterozygous germline mutations in CTLA4. Science, 2014, 345, 1623-1627.	12.6	745
2	Patients with LRBA deficiency show CTLA4 loss and immune dysregulation responsive to abatacept therapy. Science, 2015, 349, 436-440.	12.6	580
3	Dominant-activating germline mutations in the gene encoding the PI(3)K catalytic subunit p110Î´ result in T cell senescence and human immunodeficiency. Nature Immunology, 2014, 15, 88-97.	14.5	575
4	Characterization of novel stem cell factor responsive human mast cell lines LAD 1 and 2 established from a patient with mast cell sarcoma/leukemia; activation following aggregation of FcÎµRI or FcÎ³RI. Leukemia Research, 2003, 27, 677-682.	0.8	473
5	Early-onset lymphoproliferation and autoimmunity caused by germline STAT3 gain-of-function mutations. Blood, 2015, 125, 591-599.	1.4	436
6	Revised diagnostic criteria and classification for the autoimmune lymphoproliferative syndrome (ALPS): report from the 2009 NIH International Workshop. Blood, 2010, 116, e35-e40.	1.4	405
7	Heterozygous splice mutation in PIK3R1 causes human immunodeficiency with lymphoproliferation due to dominant activation of PI3K. Journal of Experimental Medicine, 2014, 211, 2537-2547.	8.5	249
8	Characterization and treatment of chronic active Epstein-Barr virus disease: a 28-year experience in the United States. Blood, 2011, 117, 5835-5849.	1.4	241
9	Effective â€œactivated PI3KÎ´ syndromeâ€â€“targeted therapy with the PI3KÎ´ inhibitor leniolisib. Blood, 2017, 130, 2307-2316.	1.4	227
10	Natural history of autoimmune lymphoproliferative syndrome associated with FAS gene mutations. Blood, 2014, 123, 1989-1999.	1.4	204
11	How I treat autoimmune lymphoproliferative syndrome. Blood, 2011, 118, 5741-5751.	1.4	156
12	Somatic KRAS mutations associated with a human nonmalignant syndrome of autoimmunity and abnormal leukocyte homeostasis. Blood, 2011, 117, 2883-2886.	1.4	139
13	Somatic FAS mutations are common in patients with genetically undefined autoimmune lymphoproliferative syndrome. Blood, 2010, 115, 5164-5169.	1.4	126
14	Use of mycophenolate mofetil for chronic, refractory immune cytopenias in children with autoimmune lymphoproliferative syndrome. British Journal of Haematology, 2005, 129, 534-538.	2.5	112
15	JMML and RALD (Ras-associated autoimmune leukoproliferative disorder): common genetic etiology yet clinically distinct entities. Blood, 2015, 125, 2753-2758.	1.4	94
16	Autoimmune Lymphoproliferative Syndrome: an Update and Review of the Literature. Current Allergy and Asthma Reports, 2014, 14, 462.	5.3	91
17	Using biomarkers to predict the presence of FAS mutations in patients with features of the autoimmune lymphoproliferative syndrome. Journal of Allergy and Clinical Immunology, 2010, 125, 946-949.e6.	2.9	87
18	FAS Inactivation Releases Unconventional Germinal Center B Cells that Escape Antigen Control and Drive IgE and Autoantibody Production. Immunity, 2015, 42, 890-902.	14.3	77

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19	Burkitt's Lymphoma: Molecular Pathogenesis and Treatment. Cancer Investigation, 2000, 18, 574-583.	1.3	75
20	Defective glycosylation and multisystem abnormalities characterize the primary immunodeficiency XMEN disease. Journal of Clinical Investigation, 2019, 130, 507-522.	8.2	74
21	Amplification of 4q21-q22 and theMXR gene in independently derived mitoxantrone-resistant cell lines. , 2000, 27, 110-116.		73
22	Use of rituximab for refractory cytopenias associated with autoimmune lymphoproliferative syndrome (ALPS). Pediatric Blood and Cancer, 2009, 52, 847-852.	1.5	70
23	Antibody responses to the SARS-CoV-2 vaccine in individuals with various inborn errors of immunity. Journal of Allergy and Clinical Immunology, 2021, 148, 1192-1197.	2.9	67
24	HEM1 deficiency disrupts mTORC2 and F-actin control in inherited immunodysregulatory disease. Science, 2020, 369, 202-207.	12.6	65
25	Inherited PD-1 deficiency underlies tuberculosis and autoimmunity in a child. Nature Medicine, 2021, 27, 1646-1654.	30.7	65
26	Abnormal B-cell maturation in the bone marrow of patients with germline mutations in PIK3CD. Journal of Allergy and Clinical Immunology, 2017, 139, 1032-1035.e6.	2.9	62
27	FAS Haploinsufficiency Is a Common Disease Mechanism in the Human Autoimmune Lymphoproliferative Syndrome. Journal of Immunology, 2011, 186, 6035-6043.	0.8	60
28	Causes and consequences of the autoimmune lymphoproliferative syndrome. Hematology, 2006, 11, 15-23.	1.5	58
29	Human PI3Kî³ deficiency and its microbiota-dependent mouse model reveal immunodeficiency and tissue immunopathology. Nature Communications, 2019, 10, 4364.	12.8	51
30	Immunodeficiency and bone marrow failure with mosaic and germline TLR8 gain of function. Blood, 2021, 137, 2450-2462.	1.4	47
31	Valproic acid (VPA), a histone deacetylase (HDAC) inhibitor, diminishes lymphoproliferation in the Fas -deficient MRL/lprâ˜'/â˜' murine model of autoimmune lymphoproliferative syndrome (ALPS). Experimental Hematology, 2009, 37, 487-494.	0.4	46
32	Development of Disseminated Histiocytic Sarcoma in a Patient With Autoimmune Lymphoproliferative Syndrome and Associated Rosai-Dorfman Disease. American Journal of Surgical Pathology, 2010, 34, 589-594.	3.7	46
33	Fas/CD95 prevents autoimmunity independently of lipid raft localization and efficient apoptosis induction. Nature Communications, 2016, 7, 13895.	12.8	45
34	Autoimmune lymphoproliferative syndrome due to FAS mutations outside the signal-transducing death domain: molecular mechanisms and clinical penetrance. Genetics in Medicine, 2012, 14, 81-89.	2.4	41
35	The autoimmune lymphoproliferative syndrome: A rare disorder providing clues about normal tolerance. Autoimmunity Reviews, 2010, 9, 488-493.	5.8	40
36	Approaches to Managing Autoimmune Cytopenias in Novel Immunological Disorders with Genetic Underpinnings Like Autoimmune Lymphoproliferative Syndrome. Frontiers in Pediatrics, 2015, 3, 65.	1.9	38

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37	Elevated interleukin-10: A new cause of dyslipidemia leading to severe HDL deficiency. Journal of Clinical Lipidology, 2015, 9, 81-90.	1.5	38
38	Biochemically deleterious human NFKB1 variants underlie an autosomal dominant form of common variable immunodeficiency. Journal of Experimental Medicine, 2021, 218, .	8.5	32
39	Characterization of ABCG2 gene amplification manifesting as extrachromosomal DNA in mitoxantrone-selected SF295 human glioblastoma cells. Cancer Genetics and Cytogenetics, 2005, 160, 126-133.	1.0	31
40	Autoimmune Lymphoproliferative Syndrome Misdiagnosed as Hemophagocytic Lymphohistiocytosis. Pediatrics, 2013, 132, e1440-e1444.	2.1	31
41	RELA haploinsufficiency in CD4 lymphoproliferative disease with autoimmune cytopenias. Journal of Allergy and Clinical Immunology, 2018, 141, 1507-1510.e8.	2.9	31
42	Fluorodeoxyglucose positron emission tomography (FDG-PET) for monitoring lymphadenopathy in the autoimmune lymphoproliferative syndrome (ALPS). American Journal of Hematology, 2006, 81, 81-85.	4.1	28
43	Elevated vitamin B12 levels in autoimmune lymphoproliferative syndrome attributable to elevated haptocorrin in lymphocytes. Clinical Biochemistry, 2012, 45, 490-492.	1.9	28
44	Up, Down, and All Around: Diagnosis and Treatment of Novel STAT3 Variant. Frontiers in Pediatrics, 2017, 5, 49.	1.9	25
45	Multibatch Cytometry Data Integration for Optimal Immunophenotyping. Journal of Immunology, 2021, 206, 206-213.	0.8	25
46	Glucose and insulin variations in patients during the time course of a FDG-PET study and implications for the â€œglucose-correctedâ€•SUV. Nuclear Medicine and Biology, 2008, 35, 441-445.	0.6	19
47	Bone marrow findings in autoimmune lymphoproliferative syndrome with germline FAS mutation. Haematologica, 2017, 102, 364-372.	3.5	19
48	Pyrimethamine treatment does not ameliorate lymphoproliferation or autoimmune disease in MRL/lprâˆ’/âˆ’ mice or in patients with autoimmune lymphoproliferative syndrome. American Journal of Hematology, 2007, 82, 1049-1055.	4.1	18
49	Paradoxical CD4 Lymphopenia in Autoimmune Lymphoproliferative Syndrome (ALPS). Frontiers in Immunology, 2019, 10, 1193.	4.8	18
50	Haploinsufficiency of immune checkpoint receptor CTLA4 induces a distinct neuroinflammatory disorder. Journal of Clinical Investigation, 2020, 130, 5551-5561.	8.2	18
51	Primary immunodeficiencies: novel genes and unusual presentations. Hematology American Society of Hematology Education Program, 2019, 2019, 443-448.	2.5	18
52	Cutting Edge: Lymphoproliferation Caused by Fas Deficiency Is Dependent on the Transcription Factor Eomesodermin. Journal of Immunology, 2010, 185, 7151-7155.	0.8	16
53	Bilateral uveitis in a patient with autoimmune lymphoproliferative syndrome. American Journal of Ophthalmology, 2005, 139, 562-563.	3.3	14
54	A Rapid Ex Vivo Clinical Diagnostic Assay for Fas Receptor-Induced T Lymphocyte Apoptosis. Journal of Clinical Immunology, 2013, 33, 479-488.	3.8	14

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55	Pulmonary Manifestations of the Autoimmune Lymphoproliferative Syndrome. A Retrospective Study of a Unique Patient Cohort. Annals of the American Thoracic Society, 2016, 13, 1279-1288.	3.2	13
56	Predispositions to Lymphoma: A Practical Review for Genetic Counselors. Journal of Genetic Counseling, 2016, 25, 1157-1170.	1.6	10
57	Management of Cytopenias in CTLA4 Haploinsufficiency Using Abatacept and Sirolimus. Blood, 2018, 132, 2409-2409.	1.4	9
58	The extent of chromosomal aberrations induced by chemotherapy in non-human primates depends on the schedule of administration. Mutation Research - Genetic Toxicology and Environmental Mutagenesis, 2005, 583, 105-119.	1.7	8
59	The contribution of rare copy number variants in FAS toward pathogenesis of autoimmune lymphoproliferative syndrome. Blood Advances, 2022, 6, 3974-3978.	5.2	8
60	In utero and early postnatal presentation of autoimmune lymphoproliferative syndrome in a family with a novel FAS mutation. Haematologica, 2013, 98, e38-e39.	3.5	7
61	Clinical Challenges: Identification of Patients With Novel Primary Immunodeficiency Syndromes. Journal of Pediatric Hematology/Oncology, 2018, 40, e319-e322.	0.6	6
62	Safety and Efficacy of Long Term Suppression of PI3Kinase Pathway By Small Molecule PI3K-Delta Inhibitor, Leniolisib in Apds (Activated PI3KÎ´ Syndrome). Blood, 2018, 132, 3706-3706.	1.4	6
63	Clinical utility gene card for: X-linked immunodeficiency with magnesium defect, Epsteinâ€“Barr virus infection, and neoplasia (XMEN). European Journal of Human Genetics, 2015, 23, 889-889.	2.8	5
64	18F-FDG PET Imaging Features of Patients With Autoimmune Lymphoproliferative Syndrome. Clinical Nuclear Medicine, 2019, 44, 949-955.	1.3	5
65	Pediatric myelodysplastic/myeloproliferative neoplasms and related diseases. Journal of Hematopathology, 2015, 8, 159-167.	0.4	4
66	Ocular Inflammatory Disorders in Autoimmune Lymphoproliferative Syndrome (ALPS). Ocular Immunology and Inflammation, 2017, 25, 708-714.	1.8	4
67	Elevated Detection of Dual Antibody B Cells Identifies Lupus Patients With B Cell-Reactive VH4-34 Autoantibodies. Frontiers in Immunology, 2022, 13, 795209.	4.8	4
68	ITP: hematologyâ€™s Cosette from Les MisÃ©rables. Blood, 2013, 121, 1928-1930.	1.4	3
69	Expanding Spectrum of Malignancies in ALPS: A Cancer Predisposing Syndrome?.. Blood, 2012, 120, 2149-2149.	1.4	3
70	Taking ALPS down a Notch. Blood, 2008, 111, 477-477.	1.4	2
71	Serendipity in splendid isolation: rapamycin. Blood, 2016, 127, 5-6.	1.4	2
72	Granulomatosis with Polyangiitis and Severe Systemic Eosinophilia Due to a Novel PIK3CD mutation. Journal of Allergy and Clinical Immunology, 2019, 143, AB425.	2.9	2

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73	Causes and Consequences of Splenectomy In ALPS-FAS. Blood, 2010, 116, 3908-3908.	1.4	2
74	Clinical Spectrum of Autoimmune Lymphoproliferative Syndrome Associated with Caspase10 Mutations (ALPS-CASP10). Blood, 2016, 128, 1335-1335.	1.4	2
75	The Role of Bone Marrow Evaluation in Clinical Allergy and Immunology Practice: When and Why. Journal of Allergy and Clinical Immunology: in Practice, 2020, 8, 3356-3362.	3.8	1
76	Clinical Spectrum of Patients with Pathogenic Variant of STAT3 conferring Gain-of-Function: A Mimic of Autoimmune Lymphoproliferative Syndrome. Blood, 2018, 132, 3723-3723.	1.4	1
77	Role of Somatic Fas Mutations in the Pathogenesis of Autoimmune Lymphoproliferative Syndrome (ALPS).. Blood, 2006, 108, 3900-3900.	1.4	1
78	Hydroxychloroquine Diminishes Lymphoproliferation in the Fas Deficient MRL/lprâ^'/â^' Murine Model of Autoimmune Lymphoproliferative Syndrome (ALPS).. Blood, 2007, 110, 1385-1385.	1.4	1
79	Indolent Phenotype of RAS-Associated Autoimmune Leukoproliferative Disorder (RALD) Is Characterized By Single Somatic Mutations in RAS Genes with Absence of Cooperating Mutations. Blood, 2016, 128, 4268-4268.	1.4	1
80	Use of Rituximab for Refractory Immune Cytopenias Associated with Autoimmune Lymphoproliferative Syndrome (ALPS).. Blood, 2007, 110, 1319-1319.	1.4	1
81	Evans syndrome: pathology and genomic hubris. Blood, 2022, 139, 312-313.	1.4	1
82	Monogenic Autoimmune Lymphoproliferative Syndromes. , 2014, , 695-709.		0
83	Early Experience with a Radiation- and Serotherapy-Free Reduced Intensity Conditioning Platform for Allogeneic Bone Marrow Transplantation (alloBMT) in Primary Immunodeficiency (PID). Biology of Blood and Marrow Transplantation, 2018, 24, S295-S297.	2.0	0
84	Spleen is cool. Pediatric Hematology and Oncology, 2019, 36, 327-329.	0.8	0
85	Whole-Body Positron Emission Tomography (PET) Using 2-18Fluoro-2-Deoxy-D-Glucose (FDG) for Monitoring Lymphadenopathy in Autoimmune Lymphoproliferative Syndrome (ALPS).. Blood, 2004, 104, 3850-3850.	1.4	0
86	Mycophenolate Mofetil and Thrombopoietin Receptor Agonists in the Treatment of Refractory Thrombocytopenia in Patients with Autoimmune Lymphoproliferative Syndrome. Blood, 2011, 118, 2218-2218.	1.4	0
87	Clinical Spectrum of RAS-Associated Autoimmune Leukoproliferative Disorder (RALD): A Distinct Clinical Entity Mimicking Juvenile Myelomonocytioc Leukemia (JMML) or Chronic Myelomonocytic Leukemia (CMML). Blood, 2012, 120, 1033-1033.	1.4	0
88	Utility of Fluorodeoxyglucose Positron Emission Tomography(FDG-PET) and Biomarkers, Serum Vitamin B12 and HDL for Assessing Lymphoproliferation in Children and Adults with Autoimmune Lymphoproliferative Syndrome Due to FAS Mutations (ALPS-FAS). Blood, 2016, 128, 4891-4891.	1.4	0
89	Spare the Spleen in ALPS: It Is Not an Expendable Vestigial Organ. Blood, 2018, 132, 2435-2435.	1.4	0
90	PI3KÎ´ inhibitor leniolisib improves symptoms in patients with APDS/PASLI. , 0, , .		0