Emmanuelle Jouanguy

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119 15,517 59 124 h-index g-index citations papers 5.46 19,119 135 15.4 L-index avg, IF ext. papers ext. citations

#	Paper	IF	Citations
119	Autoantibodies against type I IFNs in patients with life-threatening COVID-19. <i>Science</i> , 2020 , 370,	33.3	1090
118	Inborn errors of type I IFN immunity in patients with life-threatening COVID-19. Science, 2020, 370,	33.3	994
117	TLR3 deficiency in patients with herpes simplex encephalitis. <i>Science</i> , 2007 , 317, 1522-7	33.3	842
116	Interferon-gamma-receptor deficiency in an infant with fatal bacille Calmette-Gufin infection. <i>New England Journal of Medicine</i> , 1996 , 335, 1956-61	59.2	730
115	Impairment of mycobacterial immunity in human interleukin-12 receptor deficiency. <i>Science</i> , 1998 , 280, 1432-5	33.3	708
114	Impaired response to interferon-alpha/beta and lethal viral disease in human STAT1 deficiency. <i>Nature Genetics</i> , 2003 , 33, 388-91	36.3	634
113	Herpes simplex virus encephalitis in human UNC-93B deficiency. <i>Science</i> , 2006 , 314, 308-12	33.3	601
112	Gain-of-function human STAT1 mutations impair IL-17 immunity and underlie chronic mucocutaneous candidiasis. <i>Journal of Experimental Medicine</i> , 2011 , 208, 1635-48	16.6	599
111	The Jak-STAT signaling pathway is required but not sufficient for the antiviral response of drosophila. <i>Nature Immunology</i> , 2005 , 6, 946-53	19.1	466
110	A human IFNGR1 small deletion hotspot associated with dominant susceptibility to mycobacterial infection. <i>Nature Genetics</i> , 1999 , 21, 370-8	36.3	402
109	Inborn errors of IL-12/23- and IFN-gamma-mediated immunity: molecular, cellular, and clinical features. <i>Seminars in Immunology</i> , 2006 , 18, 347-61	10.7	366
108	Human intracellular ISG15 prevents interferon-Dover-amplification and auto-inflammation. <i>Nature</i> , 2015 , 517, 89-93	50.4	311
107	IL-12 and IFN-gamma in host defense against mycobacteria and salmonella in mice and men. <i>Current Opinion in Immunology</i> , 1999 , 11, 346-51	7.8	278
106	Human TRAF3 adaptor molecule deficiency leads to impaired Toll-like receptor 3 response and susceptibility to herpes simplex encephalitis. <i>Immunity</i> , 2010 , 33, 400-11	32.3	262
105	Impaired intrinsic immunity to HSV-1 in human iPSC-derived TLR3-deficient CNS cells. <i>Nature</i> , 2012 , 491, 769-73	50.4	240
104	Whole-exome sequencing-based discovery of STIM1 deficiency in a child with fatal classic Kaposi sarcoma. <i>Journal of Experimental Medicine</i> , 2010 , 207, 2307-12	16.6	236
103	Human TLR-7-, -8-, and -9-mediated induction of IFN-alpha/beta and -lambda Is IRAK-4 dependent and redundant for protective immunity to viruses. <i>Immunity</i> , 2005 , 23, 465-78	32.3	228

(2020-2011)

102	Herpes simplex encephalitis in children with autosomal recessive and dominant TRIF deficiency. Journal of Clinical Investigation, 2011 , 121, 4889-902	15.9	227
101	Herpes simplex virus encephalitis in a patient with complete TLR3 deficiency: TLR3 is otherwise redundant in protective immunity. <i>Journal of Experimental Medicine</i> , 2011 , 208, 2083-98	16.6	223
100	X-linked susceptibility to mycobacteria is caused by mutations in NEMO impairing CD40-dependent IL-12 production. <i>Journal of Experimental Medicine</i> , 2006 , 203, 1745-59	16.6	222
99	Inborn errors of interferon (IFN)-mediated immunity in humans: insights into the respective roles of IFN-alpha/beta, IFN-gamma, and IFN-lambda in host defense. <i>Immunological Reviews</i> , 2008 , 226, 29-40	11.3	220
98	Partial MCM4 deficiency in patients with growth retardation, adrenal insufficiency, and natural killer cell deficiency. <i>Journal of Clinical Investigation</i> , 2012 , 122, 821-32	15.9	201
97	IRAK-4- and MyD88-dependent pathways are essential for the removal of developing autoreactive B cells in humans. <i>Immunity</i> , 2008 , 29, 746-57	32.3	178
96	Immunological conditions of children with BCG disseminated infection. <i>Lancet, The</i> , 1995 , 346, 581	40	174
95	Human complete Stat-1 deficiency is associated with defective type I and II IFN responses in vitro but immunity to some low virulence viruses in vivo. <i>Journal of Immunology</i> , 2006 , 176, 5078-83	5.3	159
94	Partial interferon-gamma receptor signaling chain deficiency in a patient with bacille Calmette-Guffin and Mycobacterium abscessus infection. <i>Journal of Infectious Diseases</i> , 2000 , 181, 379-8	84 ⁷	152
93	A novel immunodeficiency associated with hypomorphic RAG1 mutations and CMV infection. <i>Journal of Clinical Investigation</i> , 2005 , 115, 3291-9	15.9	149
92	DOCK8 deficiency impairs CD8 T cell survival and function in humans and mice. <i>Journal of Experimental Medicine</i> , 2011 , 208, 2305-20	16.6	140
91	A partial form of recessive STAT1 deficiency in humans. <i>Journal of Clinical Investigation</i> , 2009 , 119, 1502	2 -1<u>4</u>9	140
90	Novel STAT1 alleles in otherwise healthy patients with mycobacterial disease. <i>PLoS Genetics</i> , 2006 , 2, e131	6	138
89	Human interferon-gamma-mediated immunity is a genetically controlled continuous trait that determines the outcome of mycobacterial invasion. <i>Immunological Reviews</i> , 2000 , 178, 129-37	11.3	133
88	In a novel form of IFN-gamma receptor 1 deficiency, cell surface receptors fail to bind IFN-gamma. <i>Journal of Clinical Investigation</i> , 2000 , 105, 1429-36	15.9	127
87	Recurrent staphylococcal cellulitis and subcutaneous abscesses in a child with autoantibodies against IL-6. <i>Journal of Immunology</i> , 2008 , 180, 647-54	5.3	122
86	Human Toll-like receptor-dependent induction of interferons in protective immunity to viruses. <i>Immunological Reviews</i> , 2007 , 220, 225-36	11.3	122
85	A Global Effort to Define the Human Genetics of Protective Immunity to SARS-CoV-2 Infection. <i>Cell</i> , 2020 , 181, 1194-1199	56.2	113

84	Novel primary immunodeficiencies revealed by the investigation of paediatric infectious diseases. <i>Current Opinion in Immunology</i> , 2008 , 20, 39-48	7.8	113
83	Genome-wide association study identifies variants associated with progression of liver fibrosis from HCV infection. <i>Gastroenterology</i> , 2012 , 143, 1244-1252.e12	13.3	111
82	Human RHOH deficiency causes T cell defects and susceptibility to EV-HPV infections. <i>Journal of Clinical Investigation</i> , 2012 , 122, 3239-47	15.9	109
81	TLR3 deficiency in herpes simplex encephalitis: high allelic heterogeneity and recurrence risk. <i>Neurology</i> , 2014 , 83, 1888-97	6.5	105
80	Pherokine-2 and -3. <i>FEBS Journal</i> , 2003 , 270, 3398-407		102
79	Idiopathic Disseminated Bacillus Calmette-Gue rin Infection: A French National Retrospective Study. <i>Pediatrics</i> , 1996 , 98, 774-778	7.4	102
78	Inherited MST1 deficiency underlies susceptibility to EV-HPV infections. <i>PLoS ONE</i> , 2012 , 7, e44010	3.7	101
77	Inherited human OX40 deficiency underlying classic Kaposi sarcoma of childhood. <i>Journal of Experimental Medicine</i> , 2013 , 210, 1743-59	16.6	99
76	Inborn errors of anti-viral interferon immunity in humans. Current Opinion in Virology, 2011, 1, 487-96	7.5	98
75	Mendelian susceptibility to mycobacterial infection in man. <i>Current Opinion in Immunology</i> , 1998 , 10, 413-7	7.8	92
74	Correlation of granuloma structure with clinical outcome defines two types of idiopathic disseminated BCG infection. <i>Journal of Pathology</i> , 1997 , 181, 25-30	9.4	91
73	A novel primary immunodeficiency with specific natural-killer cell deficiency maps to the centromeric region of chromosome 8. <i>American Journal of Human Genetics</i> , 2006 , 78, 721-7	11	91
72	Autoantibodies neutralizing type I IFNs are present in 4% of uninfected individuals over 70 years old and account for 20% of COVID-19 deaths. <i>Science Immunology</i> , 2021 , 6,	28	91
71	A causative relationship between mutant IFNgR1 alleles and impaired cellular response to IFNgamma in a compound heterozygous child. <i>American Journal of Human Genetics</i> , 1998 , 62, 723-6	11	90
70	Genetic susceptibility to herpes simplex virus 1 encephalitis in mice and humans. <i>Current Opinion in Allergy and Clinical Immunology</i> , 2007 , 7, 495-505	3.3	86
69	Preexisting autoantibodies to type I IFNs underlie critical COVID-19 pneumonia in patients with APS-1. <i>Journal of Experimental Medicine</i> , 2021 , 218,	16.6	79
68	Inherited IFNAR1 deficiency in otherwise healthy patients with adverse reaction to measles and yellow fever live vaccines. <i>Journal of Experimental Medicine</i> , 2019 , 216, 2057-2070	16.6	77
67	Dual T cell- and B cell-intrinsic deficiency in humans with biallelic RLTPR mutations. <i>Journal of Experimental Medicine</i> , 2016 , 213, 2413-2435	16.6	75

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66	Age-dependent Mendelian predisposition to herpes simplex virus type 1 encephalitis in childhood. <i>Journal of Pediatrics</i> , 2010 , 157, 623-9, 629.e1	3.6	75
65	Inherited GINS1 deficiency underlies growth retardation along with neutropenia and NK cell deficiency. <i>Journal of Clinical Investigation</i> , 2017 , 127, 1991-2006	15.9	73
64	X-linked recessive TLR7 deficiency in ~1% of men under 60 years old with life-threatening COVID-19. <i>Science Immunology</i> , 2021 , 6,	28	67
63	Compound heterozygous CORO1A mutations in siblings with a mucocutaneous-immunodeficiency syndrome of epidermodysplasia verruciformis-HPV, molluscum contagiosum and granulomatous tuberculoid leprosy. <i>Journal of Clinical Immunology</i> , 2014 , 34, 871-90	5.7	65
62	Osteopontin expression correlates with clinical outcome in patients with mycobacterial infection. <i>American Journal of Pathology</i> , 2000 , 157, 37-42	5.8	65
61	SARS-CoV-2 induces human plasmacytoid predendritic cell diversification via UNC93B and IRAK4. <i>Journal of Experimental Medicine</i> , 2021 , 218,	16.6	65
60	NEMO is a key component of NF- B - and IRF-3-dependent TLR3-mediated immunity to herpes simplex virus. <i>Journal of Allergy and Clinical Immunology</i> , 2011 , 128, 610-7.e1-4	11.5	57
59	Primary immunodeficiencies of protective immunity to primary infections. <i>Clinical Immunology</i> , 2010 , 135, 204-9	9	57
58	Infections in IFNGR-1-deficient children. <i>Journal of Interferon and Cytokine Research</i> , 1997 , 17, 583-7	3.5	57
57	The human CIB1-EVER1-EVER2 complex governs keratinocyte-intrinsic immunity to Epapillomaviruses. <i>Journal of Experimental Medicine</i> , 2018 , 215, 2289-2310	16.6	56
56	Human primary immunodeficiencies of type I interferons. <i>Biochimie</i> , 2007 , 89, 878-83	4.6	56
55	Homozygous gain-of-function mutation in siblings with a syndromic form of recurrent respiratory papillomatosis. <i>Proceedings of the National Academy of Sciences of the United States of America</i> , 2019 , 116, 19055-19063	11.5	55
54	Auto-antibodies to type I IFNs can underlie adverse reactions to yellow fever live attenuated vaccine. <i>Journal of Experimental Medicine</i> , 2021 , 218,	16.6	49
53	A role for interleukin-12/23 in the maturation of human natural killer and CD56+ T cells in vivo. <i>Blood</i> , 2008 , 111, 5008-16	2.2	46
52	SARS-CoV-2-related MIS-C: A key to the viral and genetic causes of Kawasaki disease?. <i>Journal of Experimental Medicine</i> , 2021 , 218,	16.6	45
51	Surface expression of the IFN-gamma R2 chain is regulated by intracellular trafficking in human T lymphocytes. <i>Journal of Immunology</i> , 2000 , 164, 201-7	5.3	44
50	Recurrent Mycobacterium avium osteomyelitis associated with a novel dominant interferon gamma receptor mutation. <i>Pediatrics</i> , 2001 , 107, E47	7.4	43
49	Inherited IL-18BP deficiency in human fulminant viral hepatitis. <i>Journal of Experimental Medicine</i> , 2019 , 216, 1777-1790	16.6	42

48	Classic Kaposi sarcoma in 3 unrelated Turkish children born to consanguineous kindreds. <i>Pediatrics</i> , 2010 , 125, e704-8	7.4	38
47	Whole-exome sequencing to analyze population structure, parental inbreeding, and familial linkage. <i>Proceedings of the National Academy of Sciences of the United States of America</i> , 2016 , 113, 671	3 ⁻¹ 8 ^{1.5}	37
46	A novel developmental and immunodeficiency syndrome associated with intrauterine growth retardation and a lack of natural killer cells. <i>Pediatrics</i> , 2004 , 113, 136-41	7.4	37
45	EVER2 deficiency is associated with mild T-cell abnormalities. <i>Journal of Clinical Immunology</i> , 2013 , 33, 14-21	5.7	33
44	Human inborn errors of immunity to herpes viruses. <i>Current Opinion in Immunology</i> , 2020 , 62, 106-122	7.8	33
43	Kaposi Sarcoma of Childhood: Inborn or Acquired Immunodeficiency to Oncogenic HHV-8. <i>Pediatric Blood and Cancer</i> , 2016 , 63, 392-7	3	33
42	JAK Inhibitor Therapy in a Child with Inherited USP18 Deficiency. <i>New England Journal of Medicine</i> , 2020 , 382, 256-265	59.2	32
41	Epidermodysplasia Verruciformis: Inborn Errors of Immunity to Human Beta-Papillomaviruses. <i>Frontiers in Microbiology</i> , 2018 , 9, 1222	5.7	32
40	Interferon-gamma receptor deficiency mimicking LangerhansRcell histiocytosis. <i>Journal of Pediatrics</i> , 2001 , 139, 600-3	3.6	31
39	Human inborn errors of immunity to infection affecting cells other than leukocytes: from the immune system to the whole organism. <i>Current Opinion in Immunology</i> , 2019 , 59, 88-100	7.8	30
38	Familial NK cell deficiency associated with impaired IL-2- and IL-15-dependent survival of lymphocytes. <i>Journal of Immunology</i> , 2006 , 177, 8835-43	5.3	29
37	A homozygous mutation of RTEL1 in a child presenting with an apparently isolated natural killer cell deficiency. <i>Journal of Allergy and Clinical Immunology</i> , 2015 , 136, 1113-4	11.5	27
36	Life-Threatening Infections Due to Live-Attenuated Vaccines: Early Manifestations of Inborn Errors of Immunity. <i>Journal of Clinical Immunology</i> , 2019 , 39, 376-390	5.7	26
35	Severe COVID-19 in the young and healthy: monogenic inborn errors of immunity?. <i>Nature Reviews Immunology</i> , 2020 , 20, 455-456	36.5	25
34	Nonpathogenic common variants of IFNGR1 and IFNGR2 in association with total serum IgE levels. Biochemical and Biophysical Research Communications, 1999 , 263, 425-9	3.4	25
33	Recurrent elevated liver transaminases and acute liver failure in two siblings with novel bi-allelic mutations of NBAS. <i>European Journal of Medical Genetics</i> , 2017 , 60, 426-432	2.6	23
32	Human genetic and immunological determinants of critical COVID-19 pneumonia Nature, 2022,	50.4	23
31	Inborn errors of the development of human natural killer cells. <i>Current Opinion in Allergy and Clinical Immunology</i> , 2013 , 13, 589-95	3.3	22

(2021-2020)

30	Fatal Cytomegalovirus Infection in an Adult with Inherited NOS2 Deficiency. <i>New England Journal of Medicine</i> , 2020 , 382, 437-445	59.2	21	
29	Requirement for both IL-12 and IFN-gamma signaling pathways in optimal IFN-gamma production by human T cells. <i>European Journal of Immunology</i> , 2002 , 32, 693-700	6.1	19	
28	TLR3 controls constitutive IFN-hantiviral immunity in human fibroblasts and cortical neurons. <i>Journal of Clinical Investigation</i> , 2021 , 131,	15.9	19	
27	Humans with inherited T´cell CD28 deficiency are susceptible to skin papillomaviruses but are otherwise healthy. <i>Cell</i> , 2021 , 184, 3812-3828.e30	56.2	18	
26	Distinct antibody repertoires against endemic human coronaviruses in children and adults. <i>JCI Insight</i> , 2021 , 6,	9.9	17	
25	Interferon gamma receptor 2 gene variants are associated with liver fibrosis in patients with chronic hepatitis C infection. <i>Gut</i> , 2010 , 59, 1120-6	19.2	15	
24	Inherited Interleukin 2-Inducible T-Cell (ITK) Kinase Deficiency in Siblings With Epidermodysplasia Verruciformis and Hodgkin Lymphoma. <i>Clinical Infectious Diseases</i> , 2019 , 68, 1938-1941	11.6	15	
23	Negative selection on human genes underlying inborn errors depends on disease outcome and both the mode and mechanism of inheritance. <i>Proceedings of the National Academy of Sciences of the United States of America</i> , 2021 , 118,	11.5	14	
22	SARS-CoV-2 induces human plasmacytoid pre-dendritic cell diversification via UNC93B and IRAK4 2021 ,		11	
21	Epidermodysplasia Verruciformis: Genetic Heterogeneity and EVER1 and EVER2 Mutations Revealed by Genome-Wide Analysis. <i>Journal of Investigative Dermatology</i> , 2019 , 139, 241-244	4.3	11	
20	A CIB1 Splice-Site Founder Mutation in Families with Typical Epidermodysplasia Verruciformis. Journal of Investigative Dermatology, 2019 , 139, 1195-1198	4.3	10	
19	TIM3+ TRBV11-2 T cells and IFNIsignature in patrolling monocytes and CD16+ NK cells delineate MIS-C <i>Journal of Experimental Medicine</i> , 2022 , 219,	16.6	9	
18	Autoantibodies Neutralizing Type I Interferons in 20% of COVID-19 Deaths in a French Hospital Journal of Clinical Immunology, 2022 , 1	5.7	7	
17	A global effort to dissect the human genetic basis of resistance to SARS-CoV-2 infection. <i>Nature Immunology</i> , 2021 ,	19.1	7	
16	Insufficient type I IFN immunity underlies life-threatening COVID-19 pneumonia. <i>Comptes Rendus - Biologies</i> , 2021 , 344, 19-25	1.4	7	
15	Identification of an Endoglin Variant Associated With HCV-Related Liver Fibrosis Progression by Next-Generation Sequencing. <i>Frontiers in Genetics</i> , 2019 , 10, 1024	4.5	6	
14	Biochemically deleterious human NFKB1 variants underlie an autosomal dominant form of common variable immunodeficiency. <i>Journal of Experimental Medicine</i> , 2021 , 218,	16.6	6	
13	Autoantibodies neutralizing type I interferons in 20% of COVID-19 deaths in a French hospital 2021		6	

12	Human genetic basis of fulminant viral hepatitis. Human Genetics, 2020, 139, 877-884	6.3	5
11	Type I interferons and SARS-CoV-2: from cells to organisms <i>Current Opinion in Immunology</i> , 2022 , 74, 172-182	7.8	4
10	Monoclonal antibody-mediated neutralization of SARS-CoV-2 in an IRF9-deficient child. <i>Proceedings of the National Academy of Sciences of the United States of America</i> , 2021 , 118,	11.5	3
9	Human genetic and immunological dissection of papillomavirus-driven diseases: new insights into their pathogenesis. <i>Current Opinion in Virology</i> , 2021 , 51, 9-15	7.5	3
8	The risk of COVID-19 death is much greater and age dependent with type I IFN autoantibodies <i>Proceedings of the National Academy of Sciences of the United States of America</i> , 2022 , 119, e22004131	19 ^{11.5}	3
7	Candidate Predisposition Variants in Kaposi Sarcoma as Detected by Whole-Genome Sequencing. Open Forum Infectious Diseases, 2019, 6, ofz337	1	1
6	. Nature Genetics, 2001 , 28, 7-9	36.3	1
65	. Nature Genetics, 2001, 28, 7-9 Negative selection on human genes causing severe inborn errors depends on disease outcome and both the mode and mechanism of inheritance	36.3	1
	Negative selection on human genes causing severe inborn errors depends on disease outcome and	36.3	
5	Negative selection on human genes causing severe inborn errors depends on disease outcome and both the mode and mechanism of inheritance		1
5	Negative selection on human genes causing severe inborn errors depends on disease outcome and both the mode and mechanism of inheritance Inherited disorders of IFN-II IFN-IIII and NF-B-mediated immunity 2013, 454-464		1