Pierluigi Gambetti

List of Publications by Year
in descending order

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140 papers	14,663 citations	22099 59 h-index	18606 119 g-index
146 all docs	146 docs citations	146 times ranked	6252 citing authors

#	Article	IF	Citations
1	Classification of sporadic Creutzfeldt-Jakob disease based on molecular and phenotypic analysis of 300 subjects. Annals of Neurology, 1999, 46, 224-233.	2.8	1,314
2	Molecular basis of phenotypic variability in sporadc creudeldt-jakob disease. Annals of Neurology, 1996, 39, 767-778.	2.8	819
3	Fatal Familial Insomnia and Dysautonomia with Selective Degeneration of Thalamic Nuclei. New England Journal of Medicine, 1986, 315, 997-1003.	13.9	688
4	Fatal familial insomnia and familial Creutzfeldt-Jakob disease: disease phenotype determined by a DNA polymorphism. Science, 1992, 258, 806-808.	6.0	658
5	Fatal Familial Insomnia, a Prion Disease with a Mutation at Codon 178 of the Prion Protein Gene. New England Journal of Medicine, 1992, 326, 444-449.	13.9	578
6	Parkinson disease, dementia, and alzheimer disease: Clinicopathological correlations. Annals of Neurology, 1980, 7, 329-335.	2.8	494
7	Truncated Forms of the Human Prion Protein in Normal Brain and in Prion Diseases. Journal of Biological Chemistry, 1995, 270, 19173-19180.	1.6	455
8	Sporadic and familial CJD: classification and characterisation. British Medical Bulletin, 2003, 66, 213-239.	2.7	449
9	Familial and sporadic fatal insomnia. Lancet Neurology, The, 2003, 2, 167-176.	4.9	321
10	Sporadic human prion diseases: molecular insights and diagnosis. Lancet Neurology, The, 2012, 11, 618-628.	4.9	319
11	Quantifying prion disease penetrance using large population control cohorts. Science Translational Medicine, 2016, 8, 322ra9.	5.8	289
12	Molecular assessment of the potential transmissibilities of BSE and scrapie to humans. Nature, 1997, 388, 285-288.	13.7	259
13	A novel human disease with abnormal prion protein sensitive to protease. Annals of Neurology, 2008, 63, 697-708.	2.8	250
14	Chronic Wasting Disease of Elk: Transmissibility to Humans Examined by Transgenic Mouse Models. Journal of Neuroscience, 2005, 25, 7944-7949.	1.7	235
15	Membrane Environment Alters the Conformational Structure of the Recombinant Human Prion Protein. Journal of Biological Chemistry, 1999, 274, 36859-36865.	1.6	230
16	Immunohistochemical localization of glial fibrillary acidic protein in human glial neoplasms. Cancer, 1980, 45, 484-494.	2.0	224
17	Aggregation and Fibrillization of the Recombinant Human Prion Protein huPrP90â”231. Biochemistry, 2000, 39, 424-431.	1.2	216
18	Variably proteaseâ€sensitive prionopathy: A new sporadic disease of the prion protein. Annals of Neurology, 2010, 68, 162-172.	2.8	203

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19	Mammalian Prions Generated from Bacterially Expressed Prion Protein in the Absence of Any Mammalian Cofactors. Journal of Biological Chemistry, 2010, 285, 14083-14087.	1.6	195
20	Fatal Familial Insomnia and Familial Creutzfeldtâ€Jakob Disease: Clinical, Pathological and Molecular Features. Brain Pathology, 1995, 5, 43-51.	2.1	192
21	Consensus classification of human prion disease histotypes allows reliable identification of molecular subtypes: an inter-rater study among surveillance centres in Europe and USA. Acta Neuropathologica, 2012, 124, 517-529.	3.9	184
22	Diagnostic and prognostic value of human prion detection in cerebrospinal fluid. Annals of Neurology, 2017, 81, 79-92.	2.8	184
23	Aberrant metal binding by prion protein in human prion disease. Journal of Neurochemistry, 2001, 78, 1400-1408.	2.1	178
24	Familial Mutations and the Thermodynamic Stability of the Recombinant Human Prion Protein. Journal of Biological Chemistry, 1998, 273, 31048-31052.	1.6	176
25	Prions in the Urine of Patients with Variant Creutzfeldtâ€“Jakob Disease. New England Journal of Medicine, 2014, 371, 530-539.	13.9	171
26	Regional distribution of protease-resistant prion protein in fatal familial insomnia. Annals of Neurology, 1995, 38, 21-29.	2.8	165
27	Cell-free propagation of prion strains. EMBO Journal, 2008, 27, 2557-2566.	3.5	164
28	Increased levels of oxidative stress markers detected in the brains of mice devoid of prion protein. Journal of Neurochemistry, 2001, 76, 565-572.	2.1	163
29	Evaluation of the Human Transmission Risk of an Atypical Bovine Spongiform Encephalopathy Prion Strain. Journal of Virology, 2008, 82, 3697-3701.	1.5	141
30	Bank Vole Prion Protein As an Apparently Universal Substrate for RT-QuIC-Based Detection and Discrimination of Prion Strains. PLoS Pathogens, 2015, 11, e1004983.	2.1	141
31	Oxidative impairment in scrapie-infected mice is associated with brain metals perturbations and altered antioxidant activities. Journal of Neurochemistry, 2008, 79, 689-698.	2.1	130
32	Identification of Novel Proteinase K-resistant C-terminal Fragments of PrP in Creutzfeldt-Jakob Disease. Journal of Biological Chemistry, 2003, 278, 40429-40436.	1.6	129
33	Classification of sporadic Creutzfeldt-Jakob disease revisited. Brain, 2006, 129, 2266-2277.	3.7	129
34	Co-existence of scrapie prion protein types 1 and 2 in sporadic Creutzfeldtâ€“Jakob disease: its effect on the phenotype and prion-type characteristics. Brain, 2009, 132, 2643-2658.	3.7	126
35	Effect of the D178N Mutation and the Codon 129 Polymorphism on the Metabolism of the Prion Protein. Journal of Biological Chemistry, 1996, 271, 12661-12668.	1.6	125
36	Abbreviated incubation times for human prions in mice expressing a chimeric mouse-human prion protein transgene. Proceedings of the National Academy of Sciences of the United States of America, 2003, 100, 4784-4789.	3.3	119

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37	Effects of Different Experimental Conditions on the PrPSc Core Generated by Protease Digestion. Journal of Biological Chemistry, 2004, 279, 16797-16804.	1.6	118
38	Heterogeneity of water-soluble amyloid Î²-peptide in Alzheimer's disease and Down's syndrome brains. FEBS Letters, 1997, 409, 411-416.	1.3	110
39	Identification of an epitope in the C terminus of normal prion protein whose expression is modulated by binding events in the N terminus 1 1Edited by F. Cohen. Journal of Molecular Biology, 2000, 301, 567-573.	2.0	110
40	Pre-symptomatic diagnosis in fatal familial insomnia: serial neurophysiological and 18FDG-PET studies. Brain, 2006, 129, 668-675.	3.7	109
41	Insoluble Aggregates and Protease-resistant Conformers of Prion Protein in Uninfected Human Brains. Journal of Biological Chemistry, 2006, 281, 34848-34858.	1.6	109
42	Antibody to DNA detects scrapie but not normal prion protein. Proceedings of the National Academy of Sciences of the United States of America, 2004, 101, 1380-1385.	3.3	103
43	Molecular Pathology of Fatal Familial Insomnia. Brain Pathology, 1998, 8, 539-548.	2.1	98
44	Molecular biology and pathology of prion strains in sporadic human prion diseases. Acta Neuropathologica, 2011, 121, 79-90.	3.9	96
45	R47H TREM2 variant increases risk of typical earlyâ€onset Alzheimer's disease but not of prion or frontotemporal dementia. Alzheimer's and Dementia, 2014, 10, 602.	0.4	94
46	The widespread alteration of neurites in Alzheimer's disease may be unrelated to amyloid deposition. Annals of Neurology, 1989, 26, 771-778.	2.8	89
47	Allelic origin of the abnormal prion protein isoform in familial prion diseases. Nature Medicine, 1997, 3, 1009-1015.	15.2	88
48	Chaperoning brain diseases. Nature, 1998, 392, 23-24.	13.7	81
49	Iatrogenic Creutzfeldt-Jakob disease with Amyloid-Î² pathology: an international study. Acta Neuropathologica Communications, 2018, 6, 5.	2.4	79
50	Alz 50 recognizes abnormal filaments in Alzheimer's disease and progressive supranuclear palsy. Annals of Neurology, 1988, 24, 407-413.	2.8	76
51	Characterization of Truncated Forms of Abnormal Prion Protein in Creutzfeldt-Jakob Disease. Journal of Biological Chemistry, 2008, 283, 30557-30565.	1.6	75
52	Amyloid Precursor Protein Metabolism in Primary Cell Cultures of Neurons, Astrocytes, and Microglia. Journal of Neurochemistry, 1996, 66, 2300-2310.	2.1	73
53	Neuronal Apoptosis in Fatal Familial Insomnia. Brain Pathology, 1998, 8, 531-537.	2.1	73
54	The Pathophysiology of Fatal Familial Insomnia. Brain Pathology, 1998, 8, 521-526.	2.1	71

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55	Amyloid fibrils from the N-terminal prion protein fragment are infectious. Proceedings of the National Academy of Sciences of the United States of America, 2016, 113, 13851-13856.	3.3	68
56	Familial spongy degeneration of the central nervous system (Van Bogaert-Bertrand disease). Acta Neuropathologica, 1969, 12, 103-115.	3.9	66
57	MORPHOLOGICAL AND BIOCHEMICAL CHANGES IN RAT SYNAPTOSOME FRACTIONS DURING NEONATAL DEVELOPMENT. Journal of Cell Biology, 1971, 51, 484-498.	2.3	66
58	THE FINE STRUCTURE OF PUROMYCIN-INDUCED CHANGES IN MOUSE ENTORHINAL CORTEX. Journal of Cell Biology, 1968, 36, 379-390.	2.3	60
59	Immunochemical Characterization of Antisera to Rat Neurofilament Subunits. Journal of Neurochemistry, 1981, 37, 1260-1265.	2.1	60
60	A refined method for molecular typing reveals that co-occurrence of PrPSc types in Creutzfeldtâ€“Jakob disease is not the rule. Laboratory Investigation, 2007, 87, 1103-1112.	1.7	60
61	Hereditary Creutzfeldt-Jakob disease and fatal familial insomnia. Clinics in Laboratory Medicine, 2003, 23, 43-64.	0.7	57
62	Multiorgan Detection and Characterization of Protease-Resistant Prion Protein in a Case of Variant CJD Examined in the United States. PLoS ONE, 2010, 5, e8765.	1.1	56
63	A novel phenotype in familial Creutzfeldt-Jakob disease: Prion protein gene E200K mutation coupled with valine at codon 129 and type 2 protease-resistant prion protein. Annals of Neurology, 1999, 45, 812-816.	2.8	55
64	Fatal familial insomnia and sporadic fatal insomnia. Handbook of Clinical Neurology / Edited By P J Vinken and G W Bruyn, 2018, 153, 271-299.	1.0	54
65	Gerstmann-StrÃussler-Scheinker disease subtypes efficiently transmit in bank voles as genuine prion diseases. Scientific Reports, 2016, 6, 20443.	1.6	54
66	Novel Differences between Two Human Prion Strains Revealed by Two-dimensional Gel Electrophoresis. Journal of Biological Chemistry, 2001, 276, 37284-37288.	1.6	53
67	Creutzfeldt-Jakob disease after liver transplantation. Annals of Neurology, 1995, 38, 269-272.	2.8	51
68	Impaired slow axonal transport in wobbler mouse motor neuron disease. Annals of Neurology, 1986, 19, 36-43.	2.8	48
69	Variably Protease-Sensitive Prionopathy: a Novel Disease of the Prion Protein. Journal of Molecular Neuroscience, 2011, 45, 422-424.	1.1	46
70	Early preclinical detection of prions in the skin of prion-infected animals. Nature Communications, 2019, 10, 247.	5.8	46
71	Evaluation of a New Criterion for Detecting Prion Disease With Diffusion Magnetic Resonance Imaging. JAMA Neurology, 2020, 77, 1141.	4.5	46
72	Influence of neuronal location on antigenic properties of neurofibrillary tangles. Annals of Neurology, 1988, 23, 604-610.	2.8	45

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73	Transmission Characteristics of Variably Protease-Sensitive Prionopathy. Emerging Infectious Diseases, 2014, 20, 2006-2014.	2.0	42
74	Identification of novel risk loci and causal insights for sporadic Creutzfeldt-Jakob disease: a genome-wide association study. Lancet Neurology, The, 2020, 19, 840-848.	4.9	42
75	Sporadic fatal insomnia: A case study. Annals of Neurology, 2000, 48, 665-669.	2.8	38
76	Spontaneous mutations in the prion protein gene causing transmissible spongiform encephalopathy. Annals of Neurology, 2002, 52, 355-359.	2.8	37
77	Small Ruminant Nor98 Prions Share Biochemical Features with Human Gerstmann-StrÃussler-Scheinker Disease and Variably Protease-Sensitive Prionopathy. PLoS ONE, 2013, 8, e66405.	1.1	37
78	Absence of protease-resistant prion protein in the cerebrospinal fluid of Creutzfeldt-Jakob disease. Journal of Pathology, 2001, 194, 9-14.	2.1	36
79	Alteration of the serotonergic nervous system in fatal familial insomnia. Annals of Neurology, 2000, 48, 788-791.	2.8	35
80	Creutzfeldt-Jakob Disease (CJD) with a Mutation at Codon 148 of Prion Protein Gene. American Journal of Pathology, 2005, 167, 1729-1738.	1.9	34
81	Gerstmann-StrÃussler-Scheinker. Journal of Neuropathology and Experimental Neurology, 2006, 65, 642-651.	0.9	33
82	Binding of Bodian's Silver and Monoclonal Antibodies to Defined Regions of Human Neurofilament Subunits: Bodian's Silver Reacts with a Highly Charged Unique Domain of Neurofilaments. Journal of Neurochemistry, 1986, 46, 366-370.	2.1	32
83	Recent US Case of Variant Creutzfeldt-Jakob Diseaseâ€"Global Implications. Emerging Infectious Diseases, 2015, 21, 750-759.	2.0	32
84	Glycoform-Selective Prion Formation in Sporadic and Familial Forms of Prion Disease. PLoS ONE, 2013, 8, e58786.	1.1	32
85	Intracerebral distribution of the abnormal isoform of the prion protein in sporadic Creutzfeldt-Jakob disease and fatal insomnia. Microscopy Research and Technique, 2000, 50, 16-25.	1.2	30
86	Subtype Diagnosis of Sporadic <scp>Creutzfeldtâ€"Jakob</scp> Disease with Diffusion <scp>Magnetic Resonance Imaging</scp>. Annals of Neurology, 2021, 89, 560-572.	2.8	30
87	Codistribution of Amyloid Î² Plaques and Spongiform Degeneration in Familial Creutzfeldt-Jakob Disease With the E200K-129M Haplotype. Archives of Neurology, 2009, 66, 1240-6.	4.9	29
88	Variably protease-sensitive prionopathy. Handbook of Clinical Neurology / Edited By P J Vinken and G W Bruyn, 2018, 153, 175-190.	1.0	29
89	The amount of slow axonal transport is proportional to the radial dimensions of the axon. Journal of Neurocytology, 1986, 15, 75-83.	1.6	28
90	Prion propagation estimated from brain diffusion MRI is subtype dependent in sporadic Creutzfeldtâ€"Jakob disease. Acta Neuropathologica, 2020, 140, 169-181.	3.9	28

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91	Familial parkinsonism and dementia with ballooned neurons, argyrophilic neuronal inclusions, atypical neurofibrillary tangles, tau-negative astrocytic fibrillary tangles, and Lewy bodies. Acta Neuropathologica, 1998, 95, 15-27.	3.9	27
92	Absence of sleep EEG markers in fatal familial insomnia healthy carriers: a spectral analysis study. Clinical Neurophysiology, 2001, 112, 1888-1892.	0.7	26
93	Fatal Familial Insomnia. Archives of Neurology, 2004, 61, 122.	4.9	25
94	Variable Protease-Sensitive Prionopathy Transmission to Bank Voles. Emerging Infectious Diseases, 2019, 25, 73-81.	2.0	25
95	Phenotypic diversity of genetic Creutzfeldtâ€“Jakob disease: a histo-molecular-based classification. Acta Neuropathologica, 2021, 142, 707-728.	3.9	24
96	Assessing Prion Infectivity of Human Urine in Sporadic Creutzfeldt-Jakob Disease. Emerging Infectious Diseases, 2012, 18, 21-28.	2.0	22
97	Distinct pathological phenotypes of Creutzfeldt-Jakob disease in recipients of prion-contaminated growth hormone. Acta Neuropathologica Communications, 2015, 3, 37.	2.4	22
98	Gerstmann-StrÃussler-Scheinker disease revisited: accumulation of covalently-linked multimers of internal prion protein fragments. Acta Neuropathologica Communications, 2019, 7, 85.	2.4	22
99	Co-existence of PrPD types 1 and 2 in sporadic Creutzfeldt-Jakob disease of the VV subgroup: phenotypic and prion protein characteristics. Scientific Reports, 2020, 10, 1503.	1.6	22
100	From Microbes to Prions. Cell, 2005, 121, 155-157.	13.5	21
101	Fluorodeoxyglucose Positron Emission Tomography (FDG-PET) Correlation of Histopathology and MRI in Prion Disease. Alzheimer Disease and Associated Disorders, 2017, 31, 1-7.	0.6	20
102	PMCA-replicated PrPD in urine of vCJD patients maintains infectivity and strain characteristics of brain PrPD: Transmission study. Scientific Reports, 2019, 9, 5191.	1.6	20
103	Functional and structural differences between the prion protein from two alleles prnpa and prnpb of mouse. FEBS Journal, 2000, 267, 2452-2459.	0.2	19
104	Failure to Detect the Presence of Prions in the Uterine and Gestational Tissues from a Gravida with Creutzfeldt-Jakob Disease. American Journal of Pathology, 2009, 174, 1602-1608.	1.9	19
105	Prions in Variably Protease-Sensitive Prionopathy: An Update. Pathogens, 2013, 2, 457-471.	1.2	19
106	Variant Creutzfeldt-Jakob Disease Death, United States. Emerging Infectious Diseases, 2005, 11, 1351-1354.	2.0	18
107	Novel strain properties distinguishing sporadic prion diseases sharing prion protein genotype and prion type. Scientific Reports, 2017, 7, 38280.	1.6	18
108	Conclusions of the Symposium. Brain Pathology, 1998, 8, 571-575.	2.1	17

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109	Comparative Study of Prions in Iatrogenic and Sporadic Creutzfeldt-Jakob Disease. Journal of Clinical & Cellular Immunology, 2014, 05, .	1.5	17
110	Protease-sensitive prions with 144-bp insertion mutations. Aging, 2013, 5, 155-173.	1.4	17
111	Role of prion protein glycosylation in replication of human prions by protein misfolding cyclic amplification. Laboratory Investigation, 2019, 99, 1741-1748.	1.7	16
112	Concealment of epitope by reduction and alkylation in prion protein. Biochemical and Biophysical Research Communications, 2005, 326, 652-659.	1.0	14
113	Characterization of the Prion Protein in Human Urine. Journal of Biological Chemistry, 2010, 285, 30489-30495.	1.6	14
114	Sporadic Fatal Insomnia Masquerading as a Paraneoplastic Cerebellar Syndrome. Archives of Neurology, 2008, 65, 971-3.	4.9	13
115	Mathematical models for the diffusion magnetic resonance signal abnormality in patients with prion diseases. NeuroImage: Clinical, 2015, 7, 142-154.	1.4	12
116	A novel mechanism of phenotypic heterogeneity in Creutzfeldt-Jakob disease. Acta Neuropathologica Communications, 2020, 8, 85.	2.4	12
117	Concurrent variably protease-sensitive prionopathy and amyotrophic lateral sclerosis. Acta Neuropathologica, 2014, 128, 313-315.	3.9	9
118	A case cluster of variant Creutzfeldt-Jakob disease linked to the Kingdom of Saudi Arabia. Brain, 2016, 139, 2609-2616.	3.7	9
119	Classification of sporadic Creutzfeldt-Jakob disease based on molecular and phenotypic analysis of 300 subjects. , 1999, 46, 224.		9
120	Co-occurrence of chronic traumatic encephalopathy and prion disease. Acta Neuropathologica Communications, 2018, 6, 140.	2.4	7
121	Impaired transmissibility of atypical prions from genetic CJDG114V. Neurology: Genetics, 2018, 4, e253.	0.9	7
122	Mechanisms of phenotypic heterogeneity in prion, Alzheimer and other conformational diseases. Journal of Alzheimer's Disease, 2001, 3, 87-95.	1.2	6
123	Clinicopathological Correlates in a <scp>PRNP</scp> P102L Mutation Carrier with Rapidly Progressing Parkinsonismâ€Dystonia. Movement Disorders Clinical Practice, 2016, 3, 355-358.	0.8	6
124	Variably Protease-sensitive Prionopathy in an Apparent Cognitively Normal 93-Year-Old. Alzheimer Disease and Associated Disorders, 2015, 29, 173-176.	0.6	6
125	Creationism and Evolutionism in Prions. American Journal of Pathology, 2013, 182, 623-627.	1.9	5
126	Two distinct conformers of PrPD type 1 of sporadic Creutzfeldtâ€“Jakob disease with codon 129VV genotype faithfully propagate in vivo. Acta Neuropathologica Communications, 2021, 9, 55.	2.4	5

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127	Phenotypic Heterogeneity of Variably Protease-Sensitive Prionopathy: A Report of Three Cases Carrying Different Genotypes at PRNP Codon 129. Viruses, 2022, 14, 367.	1.5	5
128	Human Sporadic Prion Diseases. , 2013, , 59-72.		4
129	Variants of PLCXD3 are not associated with variant or sporadic Creutzfeldt-Jakob disease in a large international study. BMC Medical Genetics, 2016, 17, 28.	2.1	3
130	A novel phenotype in familial Creutzfeldtâ€Jakob disease: Prion protein gene E200K mutation coupled with valine at codon 129 and type 2 proteaseâ€resistant prion protein. Annals of Neurology, 1999, 45, 812-816.	2.8	2
131	Alteration of the serotonergic nervous system in fatal familial insomnia. Annals of Neurology, 2000, 48, 788-791.	2.8	2
132	In Memory of Amico Bignami (1930 â€" 1994). Brain Pathology, 1995, 5, 105-107.	2.1	1
133	In Memory of Valeria Manetto (1953â€"1995). Brain Pathology, 1996, 6, 199-199.	2.1	0
134	Effect of copper on recombinant mouse prion protein. Biochemical Society Transactions, 2000, 28, A36-A36.	1.6	0
135	Tribute to Robert Terry. , 2005, 1, 83-83.		0
136	Advances in Prion Disease Surveillance. Advances in Clinical Chemistry, 2006, 41, 263-292.	1.8	0
137	Inherited mtDNA variations are not strong risk factors in human prion disease. Neurobiology of Aging, 2015, 36, 2908.e1-2908.e3.	1.5	0
138	Autobiography Series: A Life of Anecdotes. Journal of Neuropathology and Experimental Neurology, 2021, 80, 608-623.	0.9	0
139	A Novel Human Disease with Abnormal Prion Protein Sensitive to Protease. FASEB Journal, 2008, 22, 173.3.	0.2	0
140	A New Prion Disease: Protease-Sensitive Prionopathy. , 0, , 350-353.		0