Pierluigi Gambetti

List of Publications by Citations

143 papers	12,488 citations	56 h-index	111 g-index
146 ext. papers	13,715 ext. citations	10 avg, IF	5.48 L-index

#	Paper	IF	Citations
143	Classification of sporadic Creutzfeldt-Jakob disease based on molecular and phenotypic analysis of 300 subjects. *Annals of Neurology*, **1999**, 46, 224-233	9.4	1105
142	Molecular basis of phenotypic variability in sporadic Creutzfeldt-Jakob disease. *Annals of Neurology* , **1996**, 39, 767-78	9.4	705
141	Fatal familial insomnia and familial Creutzfeldt-Jakob disease: disease phenotype determined by a DNA polymorphism. *Science*, **1992**, 258, 806-8	33.3	588
140	Fatal familial insomnia and dysautonomia with selective degeneration of thalamic nuclei. *New England Journal of Medicine*, **1986**, 315, 997-1003	59.2	582
139	Fatal familial insomnia, a prion disease with a mutation at codon 178 of the prion protein gene. *New England Journal of Medicine*, **1992**, 326, 444-9	59.2	502
138	Parkinson disease, dementia, and Alzheimer disease: clinicopathological correlations. *Annals of Neurology*, **1980**, 7, 329-35	9.4	456
137	Truncated forms of the human prion protein in normal brain and in prion diseases. *Journal of Biological Chemistry*, **1995**, 270, 19173-80	5.4	401
136	Sporadic and familial CJD: classification and characterisation. *British Medical Bulletin*, **2003**, 66, 213-39	5.4	373
135	Familial and sporadic fatal insomnia. *Lancet Neurology, The*, **2003**, 2, 167-76	24.1	277
134	Sporadic human prion diseases: molecular insights and diagnosis. *Lancet Neurology, The*, **2012**, 11, 618-28	24.1	250
133	Molecular assessment of the potential transmissibilities of BSE and scrapie to humans. *Nature*, **1997**, 388, 285-8	50.4	221
132	A novel human disease with abnormal prion protein sensitive to protease. *Annals of Neurology*, **2008**, 63, 697-708	9.4	213
131	Membrane environment alters the conformational structure of the recombinant human prion protein. *Journal of Biological Chemistry*, **1999**, 274, 36859-65	5.4	213
130	Aggregation and fibrillization of the recombinant human prion protein huPrP90-231. *Biochemistry*, **2000**, 39, 424-31	3.2	210
129	Immunohistochemical localization of glial fibrillary acidic protein in human glial neoplasms. *Cancer*, **1980**, 45, 484-94	6.4	208
128	Quantifying prion disease penetrance using large population control cohorts. *Science Translational Medicine*, **2016**, 8, 322ra9	17.5	205
127	Chronic wasting disease of elk: transmissibility to humans examined by transgenic mouse models. *Journal of Neuroscience*, **2005**, 25, 7944-9	6.6	201

126	Mammalian prions generated from bacterially expressed prion protein in the absence of any mammalian cofactors. *Journal of Biological Chemistry*, **2010**, 285, 14083-7	5.4	180
125	Variably protease-sensitive prionopathy: a new sporadic disease of the prion protein. *Annals of Neurology*, **2010**, 68, 162-72	9.4	168
124	Fatal familial insomnia and familial Creutzfeldt-Jakob disease: clinical, pathological and molecular features. *Brain Pathology*, **1995**, 5, 43-51	6	167
123	Familial mutations and the thermodynamic stability of the recombinant human prion protein. *Journal of Biological Chemistry*, **1998**, 273, 31048-52	5.4	162
122	Aberrant metal binding by prion protein in human prion disease. *Journal of Neurochemistry*, **2001**, 78, 1400-8	6	154
121	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, 517-29	14.3	148
120	Regional distribution of protease-resistant prion protein in fatal familial insomnia. *Annals of Neurology*, **1995**, 38, 21-9	9.4	145
119	Prions in the urine of patients with variant Creutzfeldt-Jakob disease. *New England Journal of Medicine*, **2014**, 371, 530-9	59.2	141
118	Increased levels of oxidative stress markers detected in the brains of mice devoid of prion protein. *Journal of Neurochemistry*, **2001**, 76, 565-72	6	141
117	Cell-free propagation of prion strains. *EMBO Journal*, **2008**, 27, 2557-66	13	135
116	Diagnostic and prognostic value of human prion detection in cerebrospinal fluid. *Annals of Neurology*, **2017**, 81, 79-92	9.4	134
115	Evaluation of the human transmission risk of an atypical bovine spongiform encephalopathy prion strain. *Journal of Virology*, **2008**, 82, 3697-701	6.6	127
114	Identification of novel proteinase K-resistant C-terminal fragments of PrP in Creutzfeldt-Jakob disease. *Journal of Biological Chemistry*, **2003**, 278, 40429-36	5.4	119
113	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, 4784-9	11.5	113
112	Oxidative impairment in scrapie-infected mice is associated with brain metals perturbations and altered antioxidant activities. *Journal of Neurochemistry*, **2001**, 79, 689-98	6	112
111	Bank Vole Prion Protein As an Apparently Universal Substrate for RT-QuIC-Based Detection and Discrimination of Prion Strains. *PLoS Pathogens*, **2015**, 11, e1004983	7.6	112
110	Co-existence of scrapie prion protein types 1 and 2 in sporadic Creutzfeldt-Jakob disease: its effect on the phenotype and prion-type characteristics. *Brain*, **2009**, 132, 2643-58	11.2	111
109	Classification of sporadic Creutzfeldt-Jakob disease revisited. *Brain*, **2006**, 129, 2266-77	11.2	110

108	Effect of the D178N mutation and the codon 129 polymorphism on the metabolism of the prion protein. *Journal of Biological Chemistry*, **1996**, 271, 12661-8	5.4	110
107	Identification of an epitope in the C terminus of normal prion protein whose expression is modulated by binding events in the N terminus. *Journal of Molecular Biology*, **2000**, 301, 567-73	6.5	106
106	Effects of different experimental conditions on the PrPSc core generated by protease digestion: implications for strain typing and molecular classification of CJD. *Journal of Biological Chemistry*, **2004**, 279, 16797-804	5.4	105
105	Insoluble aggregates and protease-resistant conformers of prion protein in uninfected human brains. *Journal of Biological Chemistry*, **2006**, 281, 34848-58	5.4	96
104	Heterogeneity of water-soluble amyloid beta-peptide in Alzheimer's disease and Down's syndrome brains. *FEBS Letters*, **1997**, 409, 411-6	3.8	94
103	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, 1380-5	11.5	93
102	Pre-symptomatic diagnosis in fatal familial insomnia: serial neurophysiological and 18FDG-PET studies. *Brain*, **2006**, 129, 668-75	11.2	87
101	Molecular pathology of fatal familial insomnia. *Brain Pathology*, **1998**, 8, 539-48	6	84
100	The widespread alteration of neurites in Alzheimer's disease may be unrelated to amyloid deposition. *Annals of Neurology*, **1989**, 26, 771-8	9.4	84
99	Allelic origin of the abnormal prion protein isoform in familial prion diseases. *Nature Medicine*, **1997**, 3, 1009-15	50.5	82
98	Molecular biology and pathology of prion strains in sporadic human prion diseases. *Acta Neuropathologica*, **2011**, 121, 79-90	14.3	76
97	R47H TREM2 variant increases risk of typical early-onset Alzheimer's disease but not of prion or frontotemporal dementia. *Alzheimer's and Dementia*, **2014**, 10, 602-608.e4	1.2	74
96	Alz 50 recognizes abnormal filaments in Alzheimer's disease and progressive supranuclear palsy. *Annals of Neurology*, **1988**, 24, 407-13	9.4	71
95	Characterization of truncated forms of abnormal prion protein in Creutzfeldt-Jakob disease. *Journal of Biological Chemistry*, **2008**, 283, 30557-65	5.4	67
94	Neuronal apoptosis in fatal familial insomnia. *Brain Pathology*, **1998**, 8, 531-7	6	65
93	Morphological and biochemical changes in rat synaptosome fractions during neonatal development. *Journal of Cell Biology*, **1971**, 51, 484-98	7.3	65
92	Amyloid precursor protein metabolism in primary cell cultures of neurons, astrocytes, and microglia. *Journal of Neurochemistry*, **1996**, 66, 2300-10	6	60
91	Immunochemical characterization of antisera to rat neurofilament subunits. *Journal of Neurochemistry*, **1981**, 37, 1260-5	6	57

90	Familial spongy degeneration of the central nervous system (Van Bogaert-Bertrand disease). An ultrastructural study. *Acta Neuropathologica*, **1969**, 12, 103-15	14.3	57
89	The pathophysiology of fatal familial insomnia. *Brain Pathology*, **1998**, 8, 521-6	6	56
88	Iatrogenic Creutzfeldt-Jakob disease with Amyloid-β pathology: an international study. *Acta Neuropathologica Communications*, **2018**, 6, 5	7.3	55
87	The fine structure of puromycin-induced changes in mouse entorhinal cortex. *Journal of Cell Biology* , **1968**, 36, 379-90	7.3	55
86	Hereditary Creutzfeldt-Jakob disease and fatal familial insomnia. *Clinics in Laboratory Medicine*, **2003**, 23, 43-64	2.1	54
85	A refined method for molecular typing reveals that co-occurrence of PrP(Sc) types in Creutzfeldt-Jakob disease is not the rule. *Laboratory Investigation*, **2007**, 87, 1103-12	5.9	53
84	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	9.4	52
83	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	11.5	49
82	Impaired slow axonal transport in wobbler mouse motor neuron disease. *Annals of Neurology*, **1986**, 19, 36-43	9.4	47
81	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	3.7	46
80	Novel differences between two human prion strains revealed by two-dimensional gel electrophoresis. *Journal of Biological Chemistry*, **2001**, 276, 37284-8	5.4	45
79	Gerstmann-Sträussler-Scheinker disease subtypes efficiently transmit in bank voles as genuine prion diseases. *Scientific Reports*, **2016**, 6, 20443	4.9	44
78	Influence of neuronal location on antigenic properties of neurofibrillary tangles. *Annals of Neurology*, **1988**, 23, 604-10	9.4	42
77	Transmission characteristics of variably protease-sensitive prionopathy. *Emerging Infectious Diseases*, **2014**, 20, 2006-14	10.2	38
76	Creutzfeldt-Jakob disease after liver transplantation. *Annals of Neurology*, **1995**, 38, 269-72	9.4	37
75	Sporadic fatal insomnia: A case study. *Annals of Neurology*, **2000**, 48, 665-669	9.4	36
74	Variably protease-sensitive prionopathy: a novel disease of the prion protein. *Journal of Molecular Neuroscience*, **2011**, 45, 422-4	3.3	35
73	Small ruminant nor98 prions share biochemical features with human gerstmann-sträussler-scheinker disease and variably protease-sensitive prionopathy. *PLoS ONE*, **2013**, 8, e66405	3.7	33

72	Creutzfeldt-Jakob disease (CJD) with a mutation at codon 148 of prion protein gene: relationship with sporadic CJD. *American Journal of Pathology*, **2005**, 167, 1729-38	5.8	33
71	Absence of protease-resistant prion protein in the cerebrospinal fluid of Creutzfeldt-Jakob disease. *Journal of Pathology*, **2001**, 194, 9-14	9.4	31
70	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, 366-70	6	31
69	Early preclinical detection of prions in the skin of prion-infected animals. *Nature Communications*, **2019**, 10, 247	17.4	31
68	Gerstmann-Sträussler-Scheinker: a new phenotype with curly PrP deposits. *Journal of Neuropathology and Experimental Neurology*, **2006**, 65, 642-51	3.1	30
67	Recent US Case of Variant Creutzfeldt-Jakob Disease-Global Implications. *Emerging Infectious Diseases*, **2015**, 21, 750-9	10.2	28
66	Spontaneous mutations in the prion protein gene causing transmissible spongiform encephalopathy. *Annals of Neurology*, **2002**, 52, 355-9	9.4	28
65	Glycoform-selective prion formation in sporadic and familial forms of prion disease. *PLoS ONE*, **2013**, 8, e58786	3.7	28
64	The amount of slow axonal transport is proportional to the radial dimensions of the axon. *Journal of Neurocytology*, **1986**, 15, 75-83		27
63	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	14.3	26
62	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	2.8	26
61	Fatal familial insomnia and sporadic fatal insomnia. *Handbook of Clinical Neurology / Edited By P J Vinken and G W Bruyn*, **2018**, 153, 271-299	3	25
60	Codistribution of amyloid beta plaques and spongiform degeneration in familial Creutzfeldt-Jakob disease with the E200K-129M haplotype. *Archives of Neurology*, **2009**, 66, 1240-6		25
59	Fatal familial insomnia: the first account in a family of Chinese descent. *Archives of Neurology*, **2004**, 61, 122-5		24
58	Alteration of the serotonergic nervous system in fatal familial insomnia. *Annals of Neurology*, **2000**, 48, 788-791	9.4	24
57	Absence of sleep EEG markers in fatal familial insomnia healthy carriers: a spectral analysis study. *Clinical Neurophysiology*, **2001**, 112, 1888-92	4.3	24
56	Variably protease-sensitive prionopathy. *Handbook of Clinical Neurology / Edited By P J Vinken and G W Bruyn*, **2018**, 153, 175-190	3	20
55	Distinct pathological phenotypes of Creutzfeldt-Jakob disease in recipients of prion-contaminated growth hormone. *Acta Neuropathologica Communications*, **2015**, 3, 37	7.3	19

54	From microbes to prions the final proof of the prion hypothesis. *Cell*, **2005**, 121, 155-7	56.2	19
53	Evaluation of a New Criterion for Detecting Prion Disease With Diffusion Magnetic Resonance Imaging. *JAMA Neurology*, **2020**, 77, 1141-1149	17.2	18
52	Assessing prion infectivity of human urine in sporadic Creutzfeldt-Jakob disease. *Emerging Infectious Diseases*, **2012**, 18, 21-8	10.2	17
51	Prions in variably protease-sensitive prionopathy: an update. *Pathogens*, **2013**, 2, 457-71	4.5	17
50	Novel strain properties distinguishing sporadic prion diseases sharing prion protein genotype and prion type. *Scientific Reports*, **2017**, 7, 38280	4.9	16
49	Comparative Study of Prions in Iatrogenic and Sporadic Creutzfeldt-Jakob Disease. *Journal of Clinical & Cellular Immunology*, **2014**, 5,	2.7	16
48	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, 1602-8	5.8	16
47	Fluorodeoxyglucose Positron Emission Tomography (FDG-PET) Correlation of Histopathology and MRI in Prion Disease. *Alzheimer Disease and Associated Disorders*, **2017**, 31, 1-7	2.5	15
46	PMCA-replicated PrP in urine of vCJD patients maintains infectivity and strain characteristics of brain PrP: Transmission study. *Scientific Reports*, **2019**, 9, 5191	4.9	15
45	Functional and structural differences between the prion protein from two alleles prnp(a) and prnp(b) of mouse. *FEBS Journal*, **2000**, 267, 2452-9		15
44	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	24.1	15
43	Gerstmann-Str□ssler-Scheinker disease revisited: accumulation of covalently-linked multimers of internal prion protein fragments. *Acta Neuropathologica Communications*, **2019**, 7, 85	7.3	14
42	Concealment of epitope by reduction and alkylation in prion protein. *Biochemical and Biophysical Research Communications*, **2005**, 326, 652-9	3.4	14
41	Variant Creutzfeldt-Jakob disease death, United States. *Emerging Infectious Diseases*, **2005**, 11, 1351-4	10.2	14
40	Characterization of the prion protein in human urine. *Journal of Biological Chemistry*, **2010**, 285, 30489-95	5.4	13
39	Conclusions of the symposium. *Brain Pathology*, **1998**, 8, 571-5	6	13
38	Protease-sensitive prions with 144-bp insertion mutations. *Aging*, **2013**, 5, 155-73	5.6	13
37	Variable Protease-Sensitive Prionopathy Transmission to Bank Voles. *Emerging Infectious Diseases*, **2019**, 25, 73-81	10.2	13

36	Co-existence of PrP types 1 and 2 in sporadic Creutzfeldt-Jakob disease of the VV subgroup: phenotypic and prion protein characteristics. *Scientific Reports*, **2020**, 10, 1503	4.9	12
35	Role of prion protein glycosylation in replication of human prions by protein misfolding cyclic amplification. *Laboratory Investigation*, **2019**, 99, 1741-1748	5.9	12
34	Sporadic fatal insomnia masquerading as a paraneoplastic cerebellar syndrome. *Archives of Neurology*, **2008**, 65, 971-3		11
33	Prion propagation estimated from brain diffusion MRI is subtype dependent in sporadic Creutzfeldt-Jakob disease. *Acta Neuropathologica*, **2020**, 140, 169-181	14.3	10
32	Mathematical models for the diffusion magnetic resonance signal abnormality in patients with prion diseases. *NeuroImage: Clinical*, **2015**, 7, 142-54	5.3	10
31	Subtype Diagnosis of Sporadic Creutzfeldt-Jakob Disease with Diffusion Magnetic Resonance Imaging. *Annals of Neurology*, **2021**, 89, 560-572	9.4	10
30	Classification of sporadic Creutzfeldt-Jakob disease based on molecular and phenotypic analysis of 300 subjects **1999**, 46, 224		9
29	A novel mechanism of phenotypic heterogeneity in Creutzfeldt-Jakob disease. *Acta Neuropathologica Communications*, **2020**, 8, 85	7.3	8
28	Concurrent variably protease-sensitive prionopathy and amyotrophic lateral sclerosis. *Acta Neuropathologica*, **2014**, 128, 313-315	14.3	8
27	A case cluster of variant Creutzfeldt-Jakob disease linked to the Kingdom of Saudi Arabia. *Brain*, **2016**, 139, 2609-2616	11.2	8
26	Sporadic Creutzfeldt□Jakob Disease322-335		8
25	Impaired transmissibility of atypical prions from genetic CJD. *Neurology: Genetics*, **2018**, 4, e253	3.8	7
24	Co-occurrence of chronic traumatic encephalopathy and prion disease. *Acta Neuropathologica Communications*, **2018**, 6, 140	7.3	7
23	Creationism and evolutionism in prions. *American Journal of Pathology*, **2013**, 182, 623-7	5.8	5
22	Mechanisms of phenotypic heterogeneity in prion, Alzheimer and other conformational diseases. *Journal of Alzheimerls Disease*, **2001**, 3, 87-95	4.3	5
21	Phenotypic diversity of genetic Creutzfeldt-Jakob disease: a histo-molecular-based classification. *Acta Neuropathologica*, **2021**, 142, 707-728	14.3	5
20	Variably Protease-sensitive Prionopathy in an Apparent Cognitively Normal 93-Year-Old. *Alzheimer Disease and Associated Disorders*, **2015**, 29, 173-6	2.5	4
19	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

18	Clinicopathological Correlates in a P102L Mutation Carrier with Rapidly Progressing Parkinsonism-dystonia. *Movement Disorders Clinical Practice*, **2016**, 3, 355-358	2.2	3
17	Two distinct conformers of PrP type 1 of sporadic Creutzfeldt-Jakob disease with codon 129VV genotype faithfully propagate in vivo. *Acta Neuropathologica Communications*, **2021**, 9, 55	7.3	3
16	Genetic Creutzfeldt Jakob Disease336-345		3
15	Fatal Familial and Sporadic Insomnia346-349		3
14	Human Sporadic Prion Diseases **2013**, 59-72		3
13	Phenotypic Heterogeneity of Variably Protease-Sensitive Prionopathy: A Report of Three Cases Carrying Different Genotypes at Codon 129.. *Viruses*, **2022**, 14,	6.2	2
12	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 **1999**, 45, 812		2
11	Alteration of the serotonergic nervous system in fatal familial insomnia **2000**, 48, 788		2
10	In memory of Amico Bignami (1930-1994). *Brain Pathology*, **1995**, 5, 105-7	6	0
9	Inherited mtDNA variations are not strong risk factors in human prion disease. *Neurobiology of Aging*, **2015**, 36, 2908.e1-3	5.6	
8	Tribute to Robert Terry. *Alzheimerls and Dementia*, **2005**, 1, 83	1.2	
7	Advances in Prion Disease Surveillance. *Advances in Clinical Chemistry*, **2006**, 41, 263-292	5.8	
6	Effect of copper on recombinant mouse prion protein. *Biochemical Society Transactions*, **2000**, 28, A36-A36	3.1	
5	In Memory of Valeria Manetto (1953 1995). *Brain Pathology*, **1996**, 6, 199-199	6	
4	A Novel Human Disease with Abnormal Prion Protein Sensitive to Protease. *FASEB Journal*, **2008**, 22, 173.3	0.9	
3	The Pathology of the Synapse in Alzheimer's Disease. *Novartis Foundation Symposium*,169-183		
2	A New Prion Disease: Protease-Sensitive Prionopathy350-353		
1	Autobiography Series: A Life of Anecdotes. *Journal of Neuropathology and Experimental Neurology*, **2021**, 80, 608-623	3.1	