

Pierluigi Gambetti

List of Publications by Year in descending order

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140
papers

14,663
citations

22132
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times ranked

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citing authors

#	ARTICLE	IF	CITATIONS
1	Phenotypic Heterogeneity of Variably Protease-Sensitive Prionopathy: A Report of Three Cases Carrying Different Genotypes at PRNP Codon 129. <i>Viruses</i> , 2022, 14, 367.	1.5	5
2	Subtype Diagnosis of Sporadic <scp>Creutzfeldtâ€“Jakob</scp> Disease with Diffusion <scp>Magnetic Resonance Imaging</scp>. <i>Annals of Neurology</i> , 2021, 89, 560-572.	2.8	30
3	Two distinct conformers of PrPD type 1 of sporadic Creutzfeldtâ€“Jakob disease with codon 129VV genotype faithfully propagate in vivo. <i>Acta Neuropathologica Communications</i> , 2021, 9, 55.	2.4	5
4	Autobiography Series: A Life of Anecdotes. <i>Journal of Neuropathology and Experimental Neurology</i> , 2021, 80, 608-623.	0.9	0
5	Phenotypic diversity of genetic Creutzfeldtâ€“Jakob disease: a histo-molecular-based classification. <i>Acta Neuropathologica</i> , 2021, 142, 707-728.	3.9	24
6	Identification of novel risk loci and causal insights for sporadic Creutzfeldt-Jakob disease: a genome-wide association study. <i>Lancet Neurology</i> , The, 2020, 19, 840-848.	4.9	42
7	Evaluation of a New Criterion for Detecting Prion Disease With Diffusion Magnetic Resonance Imaging. <i>JAMA Neurology</i> , 2020, 77, 1141.	4.5	46
8	Prion propagation estimated from brain diffusion MRI is subtype dependent in sporadic Creutzfeldtâ€“Jakob disease. <i>Acta Neuropathologica</i> , 2020, 140, 169-181.	3.9	28
9	A novel mechanism of phenotypic heterogeneity in Creutzfeldt-Jakob disease. <i>Acta Neuropathologica Communications</i> , 2020, 8, 85.	2.4	12
10	Co-existence of PrPD types 1 and 2 in sporadic Creutzfeldt-Jakob disease of the VV subgroup: phenotypic and prion protein characteristics. <i>Scientific Reports</i> , 2020, 10, 1503.	1.6	22
11	Role of prion protein glycosylation in replication of human prions by protein misfolding cyclic amplification. <i>Laboratory Investigation</i> , 2019, 99, 1741-1748.	1.7	16
12	Gerstmann-Str�ussler-Scheinker disease revisited: accumulation of covalently-linked multimers of internal prion protein fragments. <i>Acta Neuropathologica Communications</i> , 2019, 7, 85.	2.4	22
13	PMCA-replicated PrPD in urine of vCJD patients maintains infectivity and strain characteristics of brain PrPD: Transmission study. <i>Scientific Reports</i> , 2019, 9, 5191.	1.6	20
14	Early preclinical detection of prions in the skin of prion-infected animals. <i>Nature Communications</i> , 2019, 10, 247.	5.8	46
15	Variable Protease-Sensitive Prionopathy Transmission to Bank Voles. <i>Emerging Infectious Diseases</i> , 2019, 25, 73-81.	2.0	25
16	Co-occurrence of chronic traumatic encephalopathy and prion disease. <i>Acta Neuropathologica Communications</i> , 2018, 6, 140.	2.4	7
17	Iatrogenic Creutzfeldt-Jakob disease with Amyloid-�2 pathology: an international study. <i>Acta Neuropathologica Communications</i> , 2018, 6, 5.	2.4	79
18	Impaired transmissibility of atypical prions from genetic CJDG114V. <i>Neurology: Genetics</i> , 2018, 4, e253.	0.9	7

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19	Variably protease-sensitive prionopathy. Handbook of Clinical Neurology / Edited By P J Vinken and G W Bruyn, 2018, 153, 175-190.	1.0	29
20	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
21	Novel strain properties distinguishing sporadic prion diseases sharing prion protein genotype and prion type. Scientific Reports, 2017, 7, 38280.	1.6	18
22	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
23	Diagnostic and prognostic value of human prion detection in cerebrospinal fluid. Annals of Neurology, 2017, 81, 79-92.	2.8	184
24	A case cluster of variant Creutzfeldt-Jakob disease linked to the Kingdom of Saudi Arabia. Brain, 2016, 139, 2609-2616.	3.7	9
25	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
26	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
27	Clinicopathological Correlates in a <i><sc>PRNP</sc></i> P102L Mutation Carrier with Rapidly Progressing Parkinsonism&Dystonia. Movement Disorders Clinical Practice, 2016, 3, 355-358.	0.8	6
28	Quantifying prion disease penetrance using large population control cohorts. Science Translational Medicine, 2016, 8, 322ra9.	5.8	289
29	Gerstmann-Str�ussler-Scheinker disease subtypes efficiently transmit in bank voles as genuine prion diseases. Scientific Reports, 2016, 6, 20443.	1.6	54
30	Mathematical models for the diffusion magnetic resonance signal abnormality in patients with prion diseases. NeuroImage: Clinical, 2015, 7, 142-154.	1.4	12
31	Inherited mtDNA variations are not strong risk factors in human prion disease. Neurobiology of Aging, 2015, 36, 2908.e1-2908.e3.	1.5	0
32	Recent US Case of Variant Creutzfeldt-Jakob Disease"Global Implications. Emerging Infectious Diseases, 2015, 21, 750-759.	2.0	32
33	Distinct pathological phenotypes of Creutzfeldt-Jakob disease in recipients of prion-contaminated growth hormone. Acta Neuropathologica Communications, 2015, 3, 37.	2.4	22
34	Variably Protease-sensitive Prionopathy in an Apparent Cognitively Normal 93-Year-Old. Alzheimer Disease and Associated Disorders, 2015, 29, 173-176.	0.6	6
35	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
36	Comparative Study of Prions in Iatrogenic and Sporadic Creutzfeldt-Jakob Disease. Journal of Clinical & Cellular Immunology, 2014, 05, .	1.5	17

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37	Transmission Characteristics of Variably Protease-Sensitive Prionopathy. Emerging Infectious Diseases, 2014, 20, 2006-2014.	2.0	42
38	<i>R47H TREM2</i> variant increases risk of typical early-onset Alzheimer's disease but not of prion or frontotemporal dementia. Alzheimer's and Dementia, 2014, 10, 602.	0.4	94
39	Prions in the Urine of Patients with Variant Creutzfeldt-Jakob Disease. New England Journal of Medicine, 2014, 371, 530-539.	13.9	171
40	Concurrent variably protease-sensitive prionopathy and amyotrophic lateral sclerosis. Acta Neuropathologica, 2014, 128, 313-315.	3.9	9
41	Creationism and Evolutionism in Prions. American Journal of Pathology, 2013, 182, 623-627.	1.9	5
42	Small Ruminant Nor98 Prions Share Biochemical Features with Human Gerstmann-Sträussler-Scheinker Disease and Variably Protease-Sensitive Prionopathy. PLoS ONE, 2013, 8, e66405.	1.1	37
43	Prions in Variably Protease-Sensitive Prionopathy: An Update. Pathogens, 2013, 2, 457-471.	1.2	19
44	Human Sporadic Prion Diseases. , 2013, , 59-72.		4
45	Glycoform-Selective Prion Formation in Sporadic and Familial Forms of Prion Disease. PLoS ONE, 2013, 8, e58786.	1.1	32
46	Protease-sensitive prions with 144-bp insertion mutations. Aging, 2013, 5, 155-173.	1.4	17
47	Sporadic human prion diseases: molecular insights and diagnosis. Lancet Neurology, The, 2012, 11, 618-628.	4.9	319
48	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
49	Assessing Prion Infectivity of Human Urine in Sporadic Creutzfeldt-Jakob Disease. Emerging Infectious Diseases, 2012, 18, 21-28.	2.0	22
50	Molecular biology and pathology of prion strains in sporadic human prion diseases. Acta Neuropathologica, 2011, 121, 79-90.	3.9	96
51	Variably Protease-Sensitive Prionopathy: a Novel Disease of the Prion Protein. Journal of Molecular Neuroscience, 2011, 45, 422-424.	1.1	46
52	Variably protease-sensitive prionopathy: A new sporadic disease of the prion protein. Annals of Neurology, 2010, 68, 162-172.	2.8	203
53	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
54	Characterization of the Prion Protein in Human Urine. Journal of Biological Chemistry, 2010, 285, 30489-30495.	1.6	14

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55	Mammalian Prions Generated from Bacterially Expressed Prion Protein in the Absence of Any Mammalian Cofactors. <i>Journal of Biological Chemistry</i> , 2010, 285, 14083-14087.	1.6	195
56	Co-existence of scrapie prion protein types 1 and 2 in sporadic Creutzfeldt-Jakob disease: its effect on the phenotype and prion-type characteristics. <i>Brain</i> , 2009, 132, 2643-2658.	3.7	126
57	Failure to Detect the Presence of Prions in the Uterine and Gestational Tissues from a Gravid with Creutzfeldt-Jakob Disease. <i>American Journal of Pathology</i> , 2009, 174, 1602-1608.	1.9	19
58	Codistribution of Amyloid β Plaques and Spongiform Degeneration in Familial Creutzfeldt-Jakob Disease With the E200K-129M Haplotype. <i>Archives of Neurology</i> , 2009, 66, 1240-6.	4.9	29
59	Oxidative impairment in scrapie-infected mice is associated with brain metals perturbations and altered antioxidant activities. <i>Journal of Neurochemistry</i> , 2008, 79, 689-698.	2.1	130
60	A novel human disease with abnormal prion protein sensitive to protease. <i>Annals of Neurology</i> , 2008, 63, 697-708.	2.8	250
61	Cell-free propagation of prion strains. <i>EMBO Journal</i> , 2008, 27, 2557-2566.	3.5	164
62	Characterization of Truncated Forms of Abnormal Prion Protein in Creutzfeldt-Jakob Disease. <i>Journal of Biological Chemistry</i> , 2008, 283, 30557-30565.	1.6	75
63	Evaluation of the Human Transmission Risk of an Atypical Bovine Spongiform Encephalopathy Prion Strain. <i>Journal of Virology</i> , 2008, 82, 3697-3701.	1.5	141
64	Sporadic Fatal Insomnia Masquerading as a Paraneoplastic Cerebellar Syndrome. <i>Archives of Neurology</i> , 2008, 65, 971-3.	4.9	13
65	A Novel Human Disease with Abnormal Prion Protein Sensitive to Protease. <i>FASEB Journal</i> , 2008, 22, 173.3.	0.2	0
66	A refined method for molecular typing reveals that co-occurrence of PrPSc types in Creutzfeldt-Jakob disease is not the rule. <i>Laboratory Investigation</i> , 2007, 87, 1103-1112.	1.7	60
67	Advances in Prion Disease Surveillance. <i>Advances in Clinical Chemistry</i> , 2006, 41, 263-292.	1.8	0
68	Gerstmann-Sträussler-Scheinker. <i>Journal of Neuropathology and Experimental Neurology</i> , 2006, 65, 642-651.	0.9	33
69	Pre-symptomatic diagnosis in fatal familial insomnia: serial neurophysiological and 18FDG-PET studies. <i>Brain</i> , 2006, 129, 668-675.	3.7	109
70	Classification of sporadic Creutzfeldt-Jakob disease revisited. <i>Brain</i> , 2006, 129, 2266-2277.	3.7	129
71	Insoluble Aggregates and Protease-resistant Conformers of Prion Protein in Uninfected Human Brains. <i>Journal of Biological Chemistry</i> , 2006, 281, 34848-34858.	1.6	109
72	Variant Creutzfeldt-Jakob Disease Death, United States. <i>Emerging Infectious Diseases</i> , 2005, 11, 1351-1354.	2.0	18

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73	Chronic Wasting Disease of Elk: Transmissibility to Humans Examined by Transgenic Mouse Models. Journal of Neuroscience, 2005, 25, 7944-7949.	1.7	235
74	Tribute to Robert Terry. , 2005, 1, 83-83.		0
75	Concealment of epitope by reduction and alkylation in prion protein. Biochemical and Biophysical Research Communications, 2005, 326, 652-659.	1.0	14
76	From Microbes to Prions. Cell, 2005, 121, 155-157.	13.5	21
77	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
78	Effects of Different Experimental Conditions on the PrP ^{Sc} Core Generated by Protease Digestion. Journal of Biological Chemistry, 2004, 279, 16797-16804.	1.6	118
79	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
80	Fatal Familial Insomnia. Archives of Neurology, 2004, 61, 122.	4.9	25
81	Familial and sporadic fatal insomnia. Lancet Neurology, The, 2003, 2, 167-176.	4.9	321
82	Hereditary Creutzfeldt-Jakob disease and fatal familial insomnia. Clinics in Laboratory Medicine, 2003, 23, 43-64.	0.7	57
83	Sporadic and familial CJD: classification and characterisation. British Medical Bulletin, 2003, 66, 213-239.	2.7	449
84	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
85	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
86	Spontaneous mutations in the prion protein gene causing transmissible spongiform encephalopathy. Annals of Neurology, 2002, 52, 355-359.	2.8	37
87	Absence of sleep EEG markers in fatal familial insomnia healthy carriers: a spectral analysis study. Clinical Neurophysiology, 2001, 112, 1888-1892.	0.7	26
88	Mechanisms of phenotypic heterogeneity in prion, Alzheimer and other conformational diseases. Journal of Alzheimer's Disease, 2001, 3, 87-95.	1.2	6
89	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
90	Aberrant metal binding by prion protein in human prion disease. Journal of Neurochemistry, 2001, 78, 1400-1408.	2.1	178

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91	Absence of protease-resistant prion protein in the cerebrospinal fluid of Creutzfeldt-Jakob disease. <i>Journal of Pathology</i> , 2001, 194, 9-14.	2.1	36
92	Novel Differences between Two Human Prion Strains Revealed by Two-dimensional Gel Electrophoresis. <i>Journal of Biological Chemistry</i> , 2001, 276, 37284-37288.	1.6	53
93	Effect of copper on recombinant mouse prion protein. <i>Biochemical Society Transactions</i> , 2000, 28, A36-A36.	1.6	0
94	Intracerebral distribution of the abnormal isoform of the prion protein in sporadic Creutzfeldt-Jakob disease and fatal insomnia. <i>Microscopy Research and Technique</i> , 2000, 50, 16-25.	1.2	30
95	Sporadic fatal insomnia: A case study. <i>Annals of Neurology</i> , 2000, 48, 665-669.	2.8	38
96	Alteration of the serotonergic nervous system in fatal familial insomnia. <i>Annals of Neurology</i> , 2000, 48, 788-791.	2.8	35
97	Functional and structural differences between the prion protein from two alleles prnpa and prnpb of mouse. <i>FEBS Journal</i> , 2000, 267, 2452-2459.	0.2	19
98	Identification of an epitope in the C terminus of normal prion protein whose expression is modulated by binding events in the N terminus 1 Edited by F. Cohen. <i>Journal of Molecular Biology</i> , 2000, 301, 567-573.	2.0	110
99	Aggregation and Fibrillization of the Recombinant Human Prion Protein huPrP90~231. <i>Biochemistry</i> , 2000, 39, 424-431.	1.2	216
100	Alteration of the serotonergic nervous system in fatal familial insomnia. <i>Annals of Neurology</i> , 2000, 48, 788-791.	2.8	2
101	Membrane Environment Alters the Conformational Structure of the Recombinant Human Prion Protein. <i>Journal of Biological Chemistry</i> , 1999, 274, 36859-36865.	1.6	230
102	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. <i>Annals of Neurology</i> , 1999, 45, 812-816.	2.8	55
103	Classification of sporadic Creutzfeldt-Jakob disease based on molecular and phenotypic analysis of 300 subjects. <i>Annals of Neurology</i> , 1999, 46, 224-233.	2.8	1,314
104	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. <i>Annals of Neurology</i> , 1999, 45, 812-816.	2.8	2
105	Classification of sporadic Creutzfeldt-Jakob disease based on molecular and phenotypic analysis of 300 subjects. , 1999, 46, 224.		9
106	Chaperoning brain diseases. <i>Nature</i> , 1998, 392, 23-24.	13.7	81
107	Familial parkinsonism and dementia with ballooned neurons, argyrophilic neuronal inclusions, atypical neurofibrillary tangles, tau-negative astrocytic fibrillary tangles, and Lewy bodies. <i>Acta Neuropathologica</i> , 1998, 95, 15-27.	3.9	27
108	Familial Mutations and the Thermodynamic Stability of the Recombinant Human Prion Protein. <i>Journal of Biological Chemistry</i> , 1998, 273, 31048-31052.	1.6	176

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109	The Pathophysiology of Fatal Familial Insomnia. Brain Pathology, 1998, 8, 521-526.	2.1	71
110	Neuronal Apoptosis in Fatal Familial Insomnia. Brain Pathology, 1998, 8, 531-537.	2.1	73
111	Molecular Pathology of Fatal Familial Insomnia. Brain Pathology, 1998, 8, 539-548.	2.1	98
112	Conclusions of the Symposium. Brain Pathology, 1998, 8, 571-575.	2.1	17
113	Heterogeneity of water-soluble amyloid β -peptide in Alzheimer's disease and Down's syndrome brains. FEBS Letters, 1997, 409, 411-416.	1.3	110
114	Allelic origin of the abnormal prion protein isoform in familial prion diseases. Nature Medicine, 1997, 3, 1009-1015.	15.2	88
115	Molecular assessment of the potential transmissibilities of BSE and scrapie to humans. Nature, 1997, 388, 285-288.	13.7	259
116	In Memory of Valeria Manetto (1953-1995). Brain Pathology, 1996, 6, 199-199.	2.1	0
117	Molecular basis of phenotypic variability in sporadic Creutzfeldt-Jakob disease. Annals of Neurology, 1996, 39, 767-778.	2.8	819
118	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
119	Amyloid Precursor Protein Metabolism in Primary Cell Cultures of Neurons, Astrocytes, and Microglia. Journal of Neurochemistry, 1996, 66, 2300-2310.	2.1	73
120	Fatal Familial Insomnia and Familial Creutzfeldt-Jakob Disease: Clinical, Pathological and Molecular Features. Brain Pathology, 1995, 5, 43-51.	2.1	192
121	In Memory of Amico Bignami (1930 - 1994). Brain Pathology, 1995, 5, 105-107.	2.1	1
122	Regional distribution of protease-resistant prion protein in fatal familial insomnia. Annals of Neurology, 1995, 38, 21-29.	2.8	165
123	Creutzfeldt-Jakob disease after liver transplantation. Annals of Neurology, 1995, 38, 269-272.	2.8	51
124	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
125	Fatal familial insomnia and familial Creutzfeldt-Jakob disease: disease phenotype determined by a DNA polymorphism. Science, 1992, 258, 806-808.	6.0	658
126	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

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127	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
128	Influence of neuronal location on antigenic properties of neurofibrillary tangles. Annals of Neurology, 1988, 23, 604-610.	2.8	45
129	Alz 50 recognizes abnormal filaments in Alzheimer's disease and progressive supranuclear palsy. Annals of Neurology, 1988, 24, 407-413.	2.8	76
130	Impaired slow axonal transport in wobbler mouse motor neuron disease. Annals of Neurology, 1986, 19, 36-43.	2.8	48
131	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
132	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
133	Fatal Familial Insomnia and Dysautonomia with Selective Degeneration of Thalamic Nuclei. New England Journal of Medicine, 1986, 315, 997-1003.	13.9	688
134	Immunochemical Characterization of Antisera to Rat Neurofilament Subunits. Journal of Neurochemistry, 1981, 37, 1260-1265.	2.1	60
135	Immunohistochemical localization of glial fibrillary acidic protein in human glial neoplasms. Cancer, 1980, 45, 484-494.	2.0	224
136	Parkinson disease, dementia, and alzheimer disease: Clinicopathological correlations. Annals of Neurology, 1980, 7, 329-335.	2.8	494
137	MORPHOLOGICAL AND BIOCHEMICAL CHANGES IN RAT SYNAPTOSOME FRACTIONS DURING NEONATAL DEVELOPMENT. Journal of Cell Biology, 1971, 51, 484-498.	2.3	66
138	Familial spongy degeneration of the central nervous system (Van Bogaert-Bertrand disease). Acta Neuropathologica, 1969, 12, 103-115.	3.9	66
139	THE FINE STRUCTURE OF PUROMYCIN-INDUCED CHANGES IN MOUSE ENTORHINAL CORTEX. Journal of Cell Biology, 1968, 36, 379-390.	2.3	60
140	A New Prion Disease: Protease-Sensitive Prionopathy. , 0, , 350-353.		0