J Paul Taylor

List of Publications by Year in descending order

145 papers	38,848 citations	7069 78 h-index	9311 143 g-index
164 all docs	164 docs citations	164 times ranked	43704 citing authors

#	Article	IF	Citations
1	Pathological phase transitions in ALS-FTD impair dynamic RNAâ€“protein granules. Rna, 2022, 28, 97-113.	1.6	15
2	Revealing the Mutational Spectrum in Southern Africans With Amyotrophic Lateral Sclerosis. Neurology: Genetics, 2022, 8, e654.	0.9	10
3	Heterozygous frameshift variants in HNRNPA2B1 cause early-onset oculopharyngeal muscular dystrophy. Nature Communications, 2022, 13, 2306.	5.8	20
4	High-fidelity reconstitution of stress granules and nucleoli in mammalian cellular lysate. Journal of Cell Biology, 2021, 220, .	2.3	56
5	TDP-43 and PINK1 mediate CHCHD10S59L mutationâ€“induced defects in Drosophila and in vitro. Nature Communications, 2021, 12, 1924.	5.8	19
6	Ubiquitination is essential for recovery of cellular activities after heat shock. Science, 2021, 372, eabc3593.	6.0	86
7	Ubiquitination of G3BP1 mediates stress granule disassembly in a context-specific manner. Science, 2021, 372, eabf6548.	6.0	151
8	Characterization of HNRNPA1 mutations defines diversity in pathogenic mechanisms and clinical presentation. JCI Insight, 2021, 6, .	2.3	38
9	Machine learning suggests polygenic risk for cognitive dysfunction in amyotrophic lateral sclerosis. EMBO Molecular Medicine, 2021, 13, e12595.	3.3	13
10	Beyond aggregation: Pathological phase transitions in neurodegenerative disease. Science, 2020, 370, 56-60.	6.0	231
11	Translational Repression of G3BP in Cancer and Germ Cells Suppresses Stress Granules and Enhances Stress Tolerance. Molecular Cell, 2020, 79, 645-659.e9.	4.5	40
12	Interactome Mapping Provides a Network of Neurodegenerative Disease Proteins and Uncovers Widespread Protein Aggregation in Affected Brains. Cell Reports, 2020, 32, 108050.	2.9	64
13	Neurotoxic microglia promote TDP-43 proteinopathy in progranulin deficiency. Nature, 2020, 588, 459-465.	13.7	98
14	hnRNPDL Phase Separation Is Regulated by Alternative Splicing and Disease-Causing Mutations Accelerate Its Aggregation. Cell Reports, 2020, 30, 1117-1128.e5.	2.9	47
15	G3BP1 Is a Tunable Switch that Triggers Phase Separation to Assemble Stress Granules. Cell, 2020, 181, 325-345.e28.	13.5	697
16	Pediatric Cancer Variant Pathogenicity Information Exchange (PeCanPIE): a cloud-based platform for curating and classifying germline variants. Genome Research, 2019, 29, 1555-1565.	2.4	28
17	Bridging biophysics and neurology: aberrant phase transitions in neurodegenerative disease. Nature Reviews Neurology, 2019, 15, 272-286.	4.9	150
18	C9orf72 Poly(PR) Dipeptide Repeats Disturb Biomolecular Phase Separation and Disrupt Nucleolar Function. Molecular Cell, 2019, 74, 713-728.e6.	4.5	128

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19	ULK1 and ULK2 Regulate Stress Granule Disassembly Through Phosphorylation and Activation of VCP/p97. Molecular Cell, 2019, 74, 742-757.e8.	4.5	123
20	Motor neuron disease-associated loss of nuclear TDP-43 is linked to DNA double-strand break repair defects. Proceedings of the National Academy of Sciences of the United States of America, 2019, 116, 4696-4705.	3.3	203
21	Rare Inherited forms of Pagetâ€™s Disease and Related Syndromes. Calcified Tissue International, 2019, 104, 501-516.	1.5	26
22	Exome sequencing in amyotrophic lateral sclerosis implicates a novel gene, DNAJC7, encoding a heat-shock protein. Nature Neuroscience, 2019, 22, 1966-1974.	7.1	101
23	Chronic optogenetic induction of stress granules is cytotoxic and reveals the evolution of ALS-FTD pathology. ELife, 2019, 8, .	2.8	184
24	Ubiquitin Modulates Liquid-Liquid Phase Separation of UBQLN2 via Disruption of Multivalent Interactions. Molecular Cell, 2018, 69, 965-978.e6.	4.5	257
25	Tau protein liquidâ€“liquid phase separation can initiate tau aggregation. EMBO Journal, 2018, 37, .	3.5	696
26	Selective modulation of the androgen receptor AF2 domain rescues degeneration in spinal bulbar muscular atrophy. Nature Medicine, 2018, 24, 427-437.	15.2	35
27	Nuclear-Import Receptors Reverse Aberrant Phase Transitions of RNA-Binding Proteins with Prion-like Domains. Cell, 2018, 173, 677-692.e20.	13.5	376
28	Stress Granule Assembly Disrupts Nucleocytoplasmic Transport. Cell, 2018, 173, 958-971.e17.	13.5	303
29	Genome-wide Analyses Identify KIF5A as a Novel ALS Gene. Neuron, 2018, 97, 1268-1283.e6.	3.8	517
30	Identification of compound heterozygous variants in OPTN in an ALS-FTD patient from the CReATe consortium: a case report. Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration, 2018, 19, 469-471.	1.1	15
31	Linking hnRNP Function to ALS and FTD Pathology. Frontiers in Neuroscience, 2018, 12, 326.	1.4	92
32	TDP-43 and RNA form amyloid-like myo-granules in regenerating muscle. Nature, 2018, 563, 508-513.	13.7	163
33	Molecular heterogeneity and CXorf67 alterations in posterior fossa group A (PFA) ependymomas. Acta Neuropathologica, 2018, 136, 211-226.	3.9	199
34	TIA1 variant drives myodegeneration in multisystem proteinopathy with SQSTM1 mutations. Journal of Clinical Investigation, 2018, 128, 1164-1177.	3.9	75
35	Effects of Mutations on the Aggregation Propensity of the Human Prion-Like Protein hnRNPA2B1. Molecular and Cellular Biology, 2017, 37, .	1.1	31
36	A PR plug for the nuclear pore in amyotrophic lateral sclerosis. Proceedings of the National Academy of Sciences of the United States of America, 2017, 114, 1445-1447.	3.3	6

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37	Therapeutic reduction of ataxin-2 extends lifespan and reduces pathology in TDP-43 mice. Nature, 2017, 544, 367-371.	13.7	422
38	Regulatory Role of RNA Chaperone TDP-43 for RNA Misfolding and Repeat-Associated Translation in SCA31. Neuron, 2017, 94, 108-124.e7.	3.8	114
39	Phase Separation of C9orf72 Dipeptide Repeats Perturbs Stress Granule Dynamics. Molecular Cell, 2017, 65, 1044-1055.e5.	4.5	437
40	Lost in Transportation: Nucleocytoplasmic Transport Defects in ALS and Other Neurodegenerative Diseases. Neuron, 2017, 96, 285-297.	3.8	208
41	TIA1 Mutations in Amyotrophic Lateral Sclerosis and Frontotemporal Dementia Promote Phase Separation and Alter Stress Granule Dynamics. Neuron, 2017, 95, 808-816.e9.	3.8	493
42	The Role of Dipeptide Repeats in C9ORF72-Related ALS-FTD. Frontiers in Molecular Neuroscience, 2017, 10, 35.	1.4	207
43	Clinical and neuropathological features of ALS/FTD with TIA1 mutations. Acta Neuropathologica Communications, 2017, 5, 96.	2.4	38
44	Sexual Reassignment Fails to Prevent Kennedyâ€™s Disease. Journal of Neuromuscular Diseases, 2016, 3, 121-125.	1.1	9
45	Higherâ€order oligomerization promotes localization of <scp>SPOP</scp> to liquid nuclear speckles. EMBO Journal, 2016, 35, 1254-1275.	3.5	172
46	Cancer-associated DDX3X mutations drive stress granule assembly and impair global translation. Scientific Reports, 2016, 6, 25996.	1.6	121
47	Decoding ALS: from genes to mechanism. Nature, 2016, 539, 197-206.	13.7	1,533
48	Targeting protein homeostasis in sporadic inclusion body myositis. Science Translational Medicine, 2016, 8, 331ra41.	5.8	99
49	C9orf72 Dipeptide Repeats Impair the Assembly, Dynamics, and Function of Membrane-Less Organelles. Cell, 2016, 167, 774-788.e17.	13.5	577
50	Protein-RNA Networks Regulated by Normal and ALS-Associated Mutant HNRNPA2B1 in the Nervous System. Neuron, 2016, 92, 780-795.	3.8	137
51	Genetic interaction of hnRNPA2B1 and DNAJB6 in aDrosophilamodel of multisystem proteinopathy. Human Molecular Genetics, 2016, 25, 936-950.	1.4	25
52	Guidelines for the use and interpretation of assays for monitoring autophagy (3rd edition). Autophagy, 2016, 12, 1-222.	4.3	4,701
53	A small-molecule Nrf1 and Nrf2 activator mitigates polyglutamine toxicity in spinal and bulbar muscular atrophy. Human Molecular Genetics, 2016, 25, 1979-1989.	1.4	55
54	Convergence of Parkin, PINK1, and Î±-Synuclein on Stress-induced Mitochondrial Morphological Remodeling. Journal of Biological Chemistry, 2015, 290, 13862-13874.	1.6	76

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55	Protein Arginine Methyltransferase 6 Enhances Polyglutamine-Expanded Androgen Receptor Function and Toxicity in Spinal and Bulbar Muscular Atrophy. Neuron, 2015, 85, 88-100.	3.8	89
56	The Role of Protein Disorder and Self-Association in the Formation of Cellular Bodies. Biophysical Journal, 2015, 108, 6a.	0.2	1
57	Network Analyses Reveal Novel Aspects of ALS Pathogenesis. PLoS Genetics, 2015, 11, e1005107.	1.5	45
58	Novel mutations expand the clinical spectrum of DYNC1H1 -associated spinal muscular atrophy. Neurology, 2015, 84, 668-679.	1.5	106
59	Phase Separation by Low Complexity Domains Promotes Stress Granule Assembly and Drives Pathological Fibrillization. Cell, 2015, 163, 123-133.	13.5	2,053
60	Multisystem proteinopathy. Neurology, 2015, 85, 658-660.	1.5	85
61	Fragile X protein mitigates TDP-43 toxicity by remodeling RNA granules and restoring translation. Human Molecular Genetics, 2015, 24, ddv389.	1.4	72
62	RAN translation at CGG repeats induces ubiquitin proteasome system impairment in models of fragile X-associated tremor ataxia syndrome. Human Molecular Genetics, 2015, 24, 4317-4326.	1.4	91
63	GGGGCC repeat expansion in C9orf72 compromises nucleocytoplasmic transport. Nature, 2015, 525, 129-133.	13.7	692
64	A case of familial ALS due to multi-system proteinopathy 1 and Huntington disease. Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration, 2015, 16, 124-126.	1.1	5
65	TDP-43 suppresses CGG repeat-induced neurotoxicity through interactions with HnRNP A2/B1. Human Molecular Genetics, 2014, 23, 5036-5051.	1.4	55
66	MFN1 deacetylation activates adaptive mitochondrial fusion and protects metabolically challenged mitochondria. Journal of Cell Science, 2014, 127, 4954-63.	1.2	91
67	G-quadruplex poses quadruple threat. Nature, 2014, 507, 175-177.	13.7	29
68	Mutations in the Matrin 3 gene cause familial amyotrophic lateral sclerosis. Nature Neuroscience, 2014, 17, 664-666.	7.1	398
69	Abnormal distribution of heterogeneous nuclear ribonucleoproteins in sporadic inclusion body myositis. Neuromuscular Disorders, 2014, 24, 611-616.	0.3	26
70	RNA metabolism in neurological disease. Brain Research, 2014, 1584, 1-2.	1.1	4
71	Profilin 1 Associates with Stress Granules and ALS-Linked Mutations Alter Stress Granule Dynamics. Journal of Neuroscience, 2014, 34, 8083-8097.	1.7	126
72	No mutations in hnRNPA1 and hnRNPA2B1 in Dutch patients with amyotrophic lateral sclerosis, frontotemporal dementia, and inclusion body myopathy. Neurobiology of Aging, 2014, 35, 1956.e9-1956.e11.	1.5	26

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73	Axonal Transport of TDP-43 mRNA Granules Is Impaired by ALS-Causing Mutations. Neuron, 2014, 81, 536-543.	3.8	521
74	Altered Ribostasis: RNA-Protein Granules in Degenerative Disorders. Cell, 2013, 154, 727-736.	13.5	543
75	VCP Is Essential for Mitochondrial Quality Control by PINK1/Parkin and this Function Is Impaired by VCP Mutations. Neuron, 2013, 78, 403.	3.8	4
76	Mutations in prion-like domains in hnRNPA2B1 and hnRNPA1 cause multisystem proteinopathy and ALS. Nature, 2013, 495, 467-473.	13.7	1,249
77	Eukaryotic Stress Granules Are Cleared by Autophagy and Cdc48/VCP Function. Cell, 2013, 153, 1461-1474.	13.5	600
78	CGG Repeat-Associated Translation Mediates Neurodegeneration in Fragile X Tremor Ataxia Syndrome. Neuron, 2013, 78, 440-455.	3.8	422
79	VCP Is Essential for Mitochondrial Quality Control by PINK1/Parkin and this Function Is Impaired by VCP Mutations. Neuron, 2013, 78, 65-80.	3.8	209
80	A functional deficiency of TERA/VCP/p97 contributes to impaired DNA repair in multiple polyglutamine diseases. Nature Communications, 2013, 4, 1816.	5.8	60
81	RNA That Gets RAN in Neurodegeneration. Science, 2013, 339, 1282-1283.	6.0	9
82	Motor neuron involvement in multisystem proteinopathy. Neurology, 2013, 80, 1874-1880.	1.5	85
83	Disease mutations in the prion-like domains of hnRNPA1 and hnRNPA2/B1 introduce potent steric zippers that drive excess RNP granule assembly. Rare Diseases (Austin, Tex), 2013, 1, e25200.	1.8	38
84	Archetypal and New Families With Alexander Disease and Novel Mutations in <emph type="ital">GFAP</emph>. Archives of Neurology, 2012, 69, 208.	4.9	28
85	Mutational analysis of the VCP gene in Parkinson's disease. Neurobiology of Aging, 2012, 33, 209.e1-209.e2.	1.5	31
86	Valosin-containing protein (VCP) mutations in sporadic amyotrophic lateral sclerosis. Neurobiology of Aging, 2012, 33, 2231.e1-2231.e6.	1.5	86
87	Guidelines for the use and interpretation of assays for monitoring autophagy. Autophagy, 2012, 8, 445-544.	4.3	3,122
88	Deficiency of ATP13A2 Leads to Lysosomal Dysfunction, Î±-Synuclein Accumulation, and Neurotoxicity. Journal of Neuroscience, 2012, 32, 4240-4246.	1.7	245
89	RNA-binding proteins in neurological disease. Brain Research, 2012, 1462, 1-2.	1.1	9
90	Huntingtin Fragments and SOD1 Mutants Form Soluble Oligomers in the Cell. PLoS ONE, 2012, 7, e40329.	1.1	5

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91	Autophagy and the Ubiquitin-Proteasome System - Protein Catabolism Comes Full Circle. , 2012, , 136-147.		0
92	Exome Sequencing Reveals VCP Mutations as a Cause of Familial ALS. Neuron, 2011, 69, 397.	3.8	7
93	Dissection and Imaging of Active Zones in the Drosophila Neuromuscular Junction. Journal of Visualized Experiments, 2011, , .	0.2	15
94	A Drosophila model of FUS-related neurodegeneration reveals genetic interaction between FUS and TDP-43. Human Molecular Genetics, 2011, 20, 2510-2523.	1.4	177
95	A Novel Conserved Isoform of the Ubiquitin Ligase UFD2a/UBE4B Is Expressed Exclusively in Mature Striated Muscle Cells. PLoS ONE, 2011, 6, e28861.	1.1	13
96	B2 attenuates polyglutamineâ€expanded androgen receptor toxicity in cell and fly models of spinal and bulbar muscular atrophy. Journal of Neuroscience Research, 2010, 88, 2207-2216.	1.3	26
97	HDAC6 controls autophagosome maturation essential for ubiquitin-selective quality-control autophagy. EMBO Journal, 2010, 29, 969-980.	3.5	660
98	Flightless flies: Drosophila models of neuromuscular disease. Annals of the New York Academy of Sciences, 2010, 1184, e1-20.	1.8	120
99	Repeat expansion disease: progress and puzzles in disease pathogenesis. Nature Reviews Genetics, 2010, 11, 247-258.	7.7	425
100	Disease mutations in Rab7 result in unregulated nucleotide exchange and inappropriate activation. Human Molecular Genetics, 2010, 19, 1033-1047.	1.4	99
101	Transgenic mice expressing mutant forms VCP/p97 recapitulate the full spectrum of IBMPFD including degeneration in muscle, brain and bone. Human Molecular Genetics, 2010, 19, 1741-1755.	1.4	171
102	Disease-causing mutations in Parkin impair mitochondrial ubiquitination, aggregation, and HDAC6-dependent mitophagy. Journal of Cell Biology, 2010, 189, 671-679.	2.3	483
103	Dynein light chain 1 is required for autophagy, protein clearance, and cell death inDrosophila. Proceedings of the National Academy of Sciences of the United States of America, 2010, 107, 742-747.	3.3	50
104	TDP-43 Mediates Degeneration in a NovelDrosophilaModel of Disease Caused by Mutations in VCP/p97. Journal of Neuroscience, 2010, 30, 7729-7739.	1.7	243
105	Histone Deacetylases Suppress CGG Repeatâ€“Induced Neurodegeneration Via Transcriptional Silencing in Models of Fragile X Tremor Ataxia Syndrome. PLoS Genetics, 2010, 6, e1001240.	1.5	93
106	VCP/p97 is essential for maturation of ubiquitin-containing autophagosomes and this function is impaired by mutations that cause IBMPFD. Autophagy, 2010, 6, 217-227.	4.3	389
107	Global Analysis of TDP-43 Interacting Proteins Reveals Strong Association with RNA Splicing and Translation Machinery. Journal of Proteome Research, 2010, 9, 1104-1120.	1.8	422
108	Native Functions of the Androgen Receptor Are Essential to Pathogenesis in a Drosophila Model of Spinobulbar Muscular Atrophy. Neuron, 2010, 67, 936-952.	3.8	150

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109	Exome Sequencing Reveals VCP Mutations as a Cause of Familial ALS. Neuron, 2010, 68, 857-864.	3.8	1,100
110	FOXO3a Is Broadly Neuroprotective In Vitro and In Vivo against Insults Implicated in Motor Neuron Diseases. Journal of Neuroscience, 2009, 29, 8236-8247.	1.7	83
111	Polyglutamine-Expanded Androgen Receptor Truncation Fragments Activate a Bax-Dependent Apoptotic Cascade Mediated by DP5/Hrk. Journal of Neuroscience, 2009, 29, 1987-1997.	1.7	56
112	Selective Accumulation of Aggregation-Prone Proteasome Substrates in Response to Proteotoxic Stress. Molecular and Cellular Biology, 2009, 29, 1774-1785.	1.1	61
113	Sarcoplasmic redistribution of nuclear TDPâ€43 in inclusion body myositis. Muscle and Nerve, 2009, 40, 19-31.	1.0	179
114	Overexpression of IGF-1 in Muscle Attenuates Disease in a Mouse Model of Spinal and Bulbar Muscular Atrophy. Neuron, 2009, 63, 316-328.	3.8	205
115	Autophagy and the ubiquitin-proteasome system: Collaborators in neuroprotection. Biochimica Et Biophysica Acta - Molecular Basis of Disease, 2008, 1782, 691-699.	1.8	302
116	The Role of Autophagy in Age-Related Neurodegeneration. NeuroSignals, 2008, 16, 75-84.	0.5	89
117	Guidelines for the use and interpretation of assays for monitoring autophagy in higher eukaryotes. Autophagy, 2008, 4, 151-175.	4.3	2,064
118	HDAC6 at the Intersection of Autophagy, the Ubiquitin-proteasome System, and Neurodegeneration. Autophagy, 2007, 3, 643-645.	4.3	107
119	Safety, Tolerability, and Pharmacokinetics of High-Dose Idebenone in Patients With Friedreich Ataxia. Archives of Neurology, 2007, 64, 803.	4.9	77
120	TDP-43 in the Ubiquitin Pathology of Frontotemporal Dementia With VCP Gene Mutations. Journal of Neuropathology and Experimental Neurology, 2007, 66, 152-157.	0.9	295
121	HDAC6 rescues neurodegeneration and provides an essential link between autophagy and the UPS. Nature, 2007, 447, 860-864.	13.7	1,068
122	Valosin-containing protein and the pathogenesis of frontotemporal dementia associated with inclusion body myopathy. Acta Neuropathologica, 2007, 114, 55-61.	3.9	56
123	Measuring Friedreich ataxia: Interrater reliability of a neurologic rating scale. Neurology, 2005, 64, 1261-1262.	1.5	316
124	Spinal Muscular Atrophy. Neurological Disease and Therapy, 2005, , 209-226.	0.0	0
125	Hsp70 dynamics in vivo: effect of heat shock and protein aggregation. Journal of Cell Science, 2004, 117, 4991-5000.	1.2	72
126	Valproic acid increases SMN levels in spinal muscular atrophy patient cells. Annals of Neurology, 2003, 54, 647-654.	2.8	269

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127	Polyglutamines Placed into Context. Neuron, 2003, 38, 681-684.	3.8	124
128	Aggresomes protect cells by enhancing the degradation of toxic polyglutamine-containing protein. Human Molecular Genetics, 2003, 12, 749-757.	1.4	378
129	A screen for drugs that protect against the cytotoxicity of polyglutamine-expanded androgen receptor. Human Molecular Genetics, 2003, 13, 437-446.	1.4	58
130	Aberrant histone acetylation, altered transcription, and retinal degeneration in a Drosophila model of polyglutamine disease are rescued by CREB-binding protein. Genes and Development, 2003, 17, 1463-1468.	2.7	130
131	Hemolytic anemia presenting as idiopathic intracranial hypertension. Neurology, 2002, 59, 960-961.	1.5	9
132	Rescue of polyglutamine-mediated cytotoxicity by double-stranded RNA-mediated RNA interference. Human Molecular Genetics, 2002, 11, 175-184.	1.4	100
133	Repeat expansion and neurological disease. , 2002, , 32-54.		4
134	Toxic Proteins in Neurodegenerative Disease. Science, 2002, 296, 1991-1995.	6.0	1,103
135	Altered acetylation in polyglutamine disease: an opportunity for therapeutic intervention?. Trends in Molecular Medicine, 2002, 8, 195-197.	3.5	17
136	Transcription of intermediate filament genes is enhanced in focal cortical dysplasia. Acta Neuropathologica, 2001, 102, 141-148.	3.9	29
137	CREB-binding protein sequestration by expanded polyglutamine. Human Molecular Genetics, 2000, 9, 2197-2202.	1.4	496
138	Activation of HIV-1 transcription by Tat in cells derived from the CNS: Evidence for the participation of NF-Î¹B â€" A review. Advances in Neuroimmunology, 1994, 4, 291-303.	1.8	22
139	Central nervous system-derived cells express a kappa B-binding activity that enhances human immunodeficiency virus type 1 transcription in vitro and facilitates TAR-independent transactivation by Tat. Journal of Virology, 1994, 68, 3971-3981.	1.5	48
140	The tumor suppressor protein p53 strongly alters human immunodeficiency virus type 1 replication. Journal of Virology, 1994, 68, 4302-4313.	1.5	86
141	TAR-Independent Activation of HIV-1 Requires the Activation Domain but Not the RNA-Binding Domain of Tat. Virology, 1993, 195, 780-785.	1.1	28
142	Activation of expression of genes coding for extracellular matrix proteins in Tat-producing glioblastoma cells.. Proceedings of the National Academy of Sciences of the United States of America, 1992, 89, 9617-9621.	3.3	101
143	Evidence that a sequence similar to TAR is important for induction of the JC virus late promoter by human immunodeficiency virus type 1 Tat. Journal of Virology, 1992, 66, 7355-7361.	1.5	68
144	TAR-independent replication of human immunodeficiency virus type 1 in glial cells. Journal of Virology, 1992, 66, 7522-7528.	1.5	46

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145	Genome-Wide Analyses Identify KIF5A as a Novel ALS Gene. SSRN Electronic Journal, 0, , .	0.4	4