J Paul Taylor

List of Publications by Year in Descending Order

148 papers	29,836 citations	72 h-index	164 g-index
164 ext. papers	34,911 ext. citations	14.6 avg, IF	6.98 L-index

#	Paper	IF	Citations
148	Revealing the Mutational Spectrum in Southern Africans With Amyotrophic Lateral Sclerosis.. *Neurology: Genetics*, **2022**, 8, e654	3.8	2
147	Heterozygous frameshift variants in HNRNPA2B1 cause early-onset oculopharyngeal muscular dystrophy.. *Nature Communications*, **2022**, 13, 2306	17.4	1
146	Pathological phase transitions in ALS-FTD impair dynamic RNA-protein granules. *Rna*, **2021**,	5.8	1
145	Machine learning suggests polygenic risk for cognitive dysfunction in amyotrophic lateral sclerosis. *EMBO Molecular Medicine*, **2021**, 13, e12595	12	3
144	TDP-43 and PINK1 mediate CHCHD10 mutation-induced defects in Drosophila and in vitro. *Nature Communications*, **2021**, 12, 1924	17.4	3
143	Ubiquitination is essential for recovery of cellular activities after heat shock. *Science*, **2021**, 372, eabc3593	33.3	20
142	Ubiquitination of G3BP1 mediates stress granule disassembly in a context-specific manner. *Science*, **2021**, 372, eabf6548	33.3	39
141	Characterization of HNRNPA1 mutations defines diversity in pathogenic mechanisms and clinical presentation. *JCI Insight*, **2021**, 6,	9.9	10
140	High-fidelity reconstitution of stress granules and nucleoli in mammalian cellular lysate. *Journal of Cell Biology*, **2021**, 220,	7.3	12
139	hnRNPDL Phase Separation Is Regulated by Alternative Splicing and Disease-Causing Mutations Accelerate Its Aggregation. *Cell Reports*, **2020**, 30, 1117-1128.e5	10.6	23
138	Beyond aggregation: Pathological phase transitions in neurodegenerative disease. *Science*, **2020**, 370, 56-60	33.3	86
137	Translational Repression of G3BP in Cancer and Germ Cells Suppresses Stress Granules and Enhances Stress Tolerance. *Molecular Cell*, **2020**, 79, 645-659.e9	17.6	11
136	Interactome Mapping Provides a Network of Neurodegenerative Disease Proteins and Uncovers Widespread Protein Aggregation in Affected Brains. *Cell Reports*, **2020**, 32, 108050	10.6	20
135	Neurotoxic microglia promote TDP-43 proteinopathy in progranulin deficiency. *Nature*, **2020**, 588, 459-465	50.4	38
134	G3BP1 Is a Tunable Switch that Triggers Phase Separation to Assemble Stress Granules. *Cell*, **2020**, 181, 325-345.e28	56.2	264
133	Pediatric Cancer Variant Pathogenicity Information Exchange (PeCanPIE): a cloud-based platform for curating and classifying germline variants. *Genome Research*, **2019**, 29, 1555-1565	9.7	16
132	Bridging biophysics and neurology: aberrant phase transitions in neurodegenerative disease. *Nature Reviews Neurology*, **2019**, 15, 272-286	15	90

131	C9orf72 Poly(PR) Dipeptide Repeats Disturb Biomolecular Phase Separation and Disrupt Nucleolar Function. *Molecular Cell*, **2019**, 74, 713-728.e6	17.6	73
130	ULK1 and ULK2 Regulate Stress Granule Disassembly Through Phosphorylation and Activation of VCP/p97. *Molecular Cell*, **2019**, 74, 742-757.e8	17.6	68
129	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	11.5	116
128	Chronic optogenetic induction of stress granules is cytotoxic and reveals the evolution of ALS-FTD pathology. *ELife*, **2019**, 8,	8.9	96
127	Author response: Chronic optogenetic induction of stress granules is cytotoxic and reveals the evolution of ALS-FTD pathology **2019**,		3
126	Rare Inherited forms of Paget's Disease and Related Syndromes. *Calcified Tissue International*, **2019**, 104, 501-516	3.9	19
125	Exome sequencing in amyotrophic lateral sclerosis implicates a novel gene, DNAJC7, encoding a heat-shock protein. *Nature Neuroscience*, **2019**, 22, 1966-1974	25.5	56
124	Ubiquitin Modulates Liquid-Liquid Phase Separation of UBQLN2 via Disruption of Multivalent Interactions. *Molecular Cell*, **2018**, 69, 965-978.e6	17.6	140
123	Tau protein liquid-liquid phase separation can initiate tau aggregation. *EMBO Journal*, **2018**, 37,	13	405
122	Selective modulation of the androgen receptor AF2 domain rescues degeneration in spinal bulbar muscular atrophy. *Nature Medicine*, **2018**, 24, 427-437	50.5	20
121	Nuclear-Import Receptors Reverse Aberrant Phase Transitions of RNA-Binding Proteins with Prion-like Domains. *Cell*, **2018**, 173, 677-692.e20	56.2	246
120	Stress Granule Assembly Disrupts Nucleocytoplasmic Transport. *Cell*, **2018**, 173, 958-971.e17	56.2	195
119	Genome-wide Analyses Identify KIF5A as a Novel ALS Gene. *Neuron*, **2018**, 97, 1268-1283.e6	13.9	296
118	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	3.6	12
117	Molecular heterogeneity and CXorf67 alterations in posterior fossa group A (PFA) ependymomas. *Acta Neuropathologica*, **2018**, 136, 211-226	14.3	111
116	TIA1 variant drives myodegeneration in multisystem proteinopathy with SQSTM1 mutations. *Journal of Clinical Investigation*, **2018**, 128, 1164-1177	15.9	53
115	Linking hnRNP Function to ALS and FTD Pathology. *Frontiers in Neuroscience*, **2018**, 12, 326	5.1	60
114	TDP-43 and RNA form amyloid-like myo-granules in regenerating muscle. *Nature*, **2018**, 563, 508-513	50.4	104

113	Effects of Mutations on the Aggregation Propensity of the Human Prion-Like Protein hnRNPA2B1. *Molecular and Cellular Biology*, **2017**, 37,	4.8	26
112	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	11.5	5
111	Therapeutic reduction of ataxin-2 extends lifespan and reduces pathology in TDP-43 mice. *Nature*, **2017**, 544, 367-371	50.4	278
110	Regulatory Role of RNA Chaperone TDP-43 for RNA Misfolding and Repeat-Associated Translation in SCA31. *Neuron*, **2017**, 94, 108-124.e7	13.9	78
109	Phase Separation of C9orf72 Dipeptide Repeats Perturbs Stress Granule Dynamics. *Molecular Cell*, **2017**, 65, 1044-1055.e5	17.6	307
108	Lost in Transportation: Nucleocytoplasmic Transport Defects in ALS and Other Neurodegenerative Diseases. *Neuron*, **2017**, 96, 285-297	13.9	154
107	Clinical and neuropathological features of ALS/FTD with TIA1 mutations. *Acta Neuropathologica Communications*, **2017**, 5, 96	7.3	27
106	TIA1 Mutations in Amyotrophic Lateral Sclerosis and Frontotemporal Dementia Promote Phase Separation and Alter Stress Granule Dynamics. *Neuron*, **2017**, 95, 808-816.e9	13.9	341
105	The Role of Dipeptide Repeats in C9ORF72-Related ALS-FTD. *Frontiers in Molecular Neuroscience*, **2017**, 10, 35	6.1	132
104	Decoding ALS: from genes to mechanism. *Nature*, **2016**, 539, 197-206	50.4	1040
103	Targeting protein homeostasis in sporadic inclusion body myositis. *Science Translational Medicine*, **2016**, 8, 331ra41	17.5	69
102	C9orf72 Dipeptide Repeats Impair the Assembly, Dynamics, and Function of Membrane-Less Organelles. *Cell*, **2016**, 167, 774-788.e17	56.2	402
101	Protein-RNA Networks Regulated by Normal and ALS-Associated Mutant HNRNPA2B1 in the Nervous System. *Neuron*, **2016**, 92, 780-795	13.9	94
100	Genetic interaction of hnRNPA2B1 and DNAJB6 in a Drosophila model of multisystem proteinopathy. *Human Molecular Genetics*, **2016**, 25, 936-50	5.6	21
99	Guidelines for the use and interpretation of assays for monitoring autophagy (3rd edition). *Autophagy*, **2016**, 12, 1-222	10.2	3838
98	A small-molecule Nrf1 and Nrf2 activator mitigates polyglutamine toxicity in spinal and bulbar muscular atrophy. *Human Molecular Genetics*, **2016**, 25, 1979-1989	5.6	34
97	Sexual Reassignment Fails to Prevent Kennedy's Disease. *Journal of Neuromuscular Diseases*, **2016**, 3, 121-125	5	6
96	Higher-order oligomerization promotes localization of SPOP to liquid nuclear speckles. *EMBO Journal*, **2016**, 35, 1254-75	13	113

95	Cancer-associated DDX3X mutations drive stress granule assembly and impair global translation. *Scientific Reports*, **2016**, 6, 25996	4.9	75
94	Network analyses reveal novel aspects of ALS pathogenesis. *PLoS Genetics*, **2015**, 11, e1005107	6	36
93	Novel mutations expand the clinical spectrum of DYNC1H1-associated spinal muscular atrophy. *Neurology*, **2015**, 84, 668-79	6.5	81
92	Phase separation by low complexity domains promotes stress granule assembly and drives pathological fibrillization. *Cell*, **2015**, 163, 123-33	56.2	1339
91	Fragile X protein mitigates TDP-43 toxicity by remodeling RNA granules and restoring translation. *Human Molecular Genetics*, **2015**, 24, 6886-98	5.6	59
90	RAN translation at CGG repeats induces ubiquitin proteasome system impairment in models of fragile X-associated tremor ataxia syndrome. *Human Molecular Genetics*, **2015**, 24, 4317-26	5.6	73
89	GGGGCC repeat expansion in C9orf72 compromises nucleocytoplasmic transport. *Nature*, **2015**, 525, 129-33	50.4	540
88	A case of familial ALS due to multi-system proteinopathy 1 and Huntington disease. *Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration*, **2015**, 16, 124-6	3.6	3
87	Convergence of Parkin, PINK1, and Synuclein on Stress-induced Mitochondrial Morphological Remodeling. *Journal of Biological Chemistry*, **2015**, 290, 13862-74	5.4	59
86	Protein arginine methyltransferase 6 enhances polyglutamine-expanded androgen receptor function and toxicity in spinal and bulbar muscular atrophy. *Neuron*, **2015**, 85, 88-100	13.9	62
85	Mutations in the Matrin 3 gene cause familial amyotrophic lateral sclerosis. *Nature Neuroscience*, **2014**, 17, 664-666	25.5	319
84	Abnormal distribution of heterogeneous nuclear ribonucleoproteins in sporadic inclusion body myositis. *Neuromuscular Disorders*, **2014**, 24, 611-6	2.9	22
83	Profilin 1 associates with stress granules and ALS-linked mutations alter stress granule dynamics. *Journal of Neuroscience*, **2014**, 34, 8083-97	6.6	102
82	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	5.6	20
81	Axonal transport of TDP-43 mRNA granules is impaired by ALS-causing mutations. *Neuron*, **2014**, 81, 536-543	13.9	408
80	TDP-43 suppresses CGG repeat-induced neurotoxicity through interactions with HnRNP A2/B1. *Human Molecular Genetics*, **2014**, 23, 5036-51	5.6	49
79	MFN1 deacetylation activates adaptive mitochondrial fusion and protects metabolically challenged mitochondria. *Journal of Cell Science*, **2014**, 127, 4954-63	5.3	71
78	Neurodegenerative diseases: G-quadruplex poses quadruple threat. *Nature*, **2014**, 507, 175-7	50.4	26

77	Altered ribostasis: RNA-protein granules in degenerative disorders. *Cell*, **2013**, 154, 727-36	56.2	423
76	VCP Is Essential for Mitochondrial Quality Control by PINK1/Parkin and this Function Is Impaired by VCP Mutations. *Neuron*, **2013**, 78, 403	13.9	3
75	Mutations in prion-like domains in hnRNPA2B1 and hnRNPA1 cause multisystem proteinopathy and ALS. *Nature*, **2013**, 495, 467-73	50.4	965
74	Eukaryotic stress granules are cleared by autophagy and Cdc48/VCP function. *Cell*, **2013**, 153, 1461-74	56.2	457
73	CGG repeat-associated translation mediates neurodegeneration in fragile X tremor ataxia syndrome. *Neuron*, **2013**, 78, 440-55	13.9	327
72	VCP is essential for mitochondrial quality control by PINK1/Parkin and this function is impaired by VCP mutations. *Neuron*, **2013**, 78, 65-80	13.9	170
71	A functional deficiency of TERA/VCP/p97 contributes to impaired DNA repair in multiple polyglutamine diseases. *Nature Communications*, **2013**, 4, 1816	17.4	49
70	Neuroscience. RNA that gets RAN in neurodegeneration. *Science*, **2013**, 339, 1282-3	33.3	8
69	Motor neuron involvement in multisystem proteinopathy: implications for ALS. *Neurology*, **2013**, 80, 1874-80	6.9	68
68	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		30
67	Mutational analysis of the VCP gene in Parkinson□s disease. *Neurobiology of Aging*, **2012**, 33, 209.e1-2	5.6	24
66	Valosin-containing protein (VCP) mutations in sporadic amyotrophic lateral sclerosis. *Neurobiology of Aging*, **2012**, 33, 2231.e1-2231.e6	5.6	74
65	Guidelines for the use and interpretation of assays for monitoring autophagy. *Autophagy*, **2012**, 8, 445-544	10.2	2783
64	Deficiency of ATP13A2 leads to lysosomal dysfunction, □synuclein accumulation, and neurotoxicity. *Journal of Neuroscience*, **2012**, 32, 4240-6	6.6	212
63	Archetypal and new families with Alexander disease and novel mutations in GFAP. *Archives of Neurology*, **2012**, 69, 208-14		22
62	Huntingtin fragments and SOD1 mutants form soluble oligomers in the cell. *PLoS ONE*, **2012**, 7, e40329	3.7	5
61	Exome Sequencing Reveals VCP Mutations as a Cause of Familial ALS. *Neuron*, **2011**, 69, 397	13.9	4
60	Dissection and imaging of active zones in the Drosophila neuromuscular junction. *Journal of Visualized Experiments*, **2011**,	1.6	14

59	A Drosophila model of FUS-related neurodegeneration reveals genetic interaction between FUS and TDP-43. *Human Molecular Genetics*, **2011**, 20, 2510-23	5.6	153
58	A novel conserved isoform of the ubiquitin ligase UFD2a/UBE4B is expressed exclusively in mature striated muscle cells. *PLoS ONE*, **2011**, 6, e28861	3.7	9
57	HDAC6 controls autophagosome maturation essential for ubiquitin-selective quality-control autophagy. *EMBO Journal*, **2010**, 29, 969-80	13	584
56	Flightless flies: Drosophila models of neuromuscular disease. *Annals of the New York Academy of Sciences*, **2010**, 1184, e1-20	6.5	98
55	Repeat expansion disease: progress and puzzles in disease pathogenesis. *Nature Reviews Genetics*, **2010**, 11, 247-58	30.1	341
54	Disease mutations in Rab7 result in unregulated nucleotide exchange and inappropriate activation. *Human Molecular Genetics*, **2010**, 19, 1033-47	5.6	79
53	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, 1741-55	5.6	138
52	Disease-causing mutations in parkin impair mitochondrial ubiquitination, aggregation, and HDAC6-dependent mitophagy. *Journal of Cell Biology*, **2010**, 189, 671-9	7.3	420
51	Dynein light chain 1 is required for autophagy, protein clearance, and cell death in Drosophila. *Proceedings of the National Academy of Sciences of the United States of America*, **2010**, 107, 742-7	11.5	43
50	TDP-43 mediates degeneration in a novel Drosophila model of disease caused by mutations in VCP/p97. *Journal of Neuroscience*, **2010**, 30, 7729-39	6.6	204
49	Histone deacetylases suppress CGG repeat-induced neurodegeneration via transcriptional silencing in models of fragile X tremor ataxia syndrome. *PLoS Genetics*, **2010**, 6, e1001240	6	83
48	VCP/p97 is essential for maturation of ubiquitin-containing autophagosomes and this function is impaired by mutations that cause IBMPFD. *Autophagy*, **2010**, 6, 217-27	10.2	339
47	Global analysis of TDP-43 interacting proteins reveals strong association with RNA splicing and translation machinery. *Journal of Proteome Research*, **2010**, 9, 1104-20	5.6	328
46	Native functions of the androgen receptor are essential to pathogenesis in a Drosophila model of spinobulbar muscular atrophy. *Neuron*, **2010**, 67, 936-52	13.9	132
45	Exome sequencing reveals VCP mutations as a cause of familial ALS. *Neuron*, **2010**, 68, 857-64	13.9	939
44	B2 attenuates polyglutamine-expanded androgen receptor toxicity in cell and fly models of spinal and bulbar muscular atrophy. *Journal of Neuroscience Research*, **2010**, 88, 2207-16	4.4	22
43	FOXO3a is broadly neuroprotective in vitro and in vivo against insults implicated in motor neuron diseases. *Journal of Neuroscience*, **2009**, 29, 8236-47	6.6	72
42	Polyglutamine-expanded androgen receptor truncation fragments activate a Bax-dependent apoptotic cascade mediated by DP5/Hrk. *Journal of Neuroscience*, **2009**, 29, 1987-97	6.6	51

41	Selective accumulation of aggregation-prone proteasome substrates in response to proteotoxic stress. *Molecular and Cellular Biology*, **2009**, 29, 1774-85	4.8	48
40	Sarcoplasmic redistribution of nuclear TDP-43 in inclusion body myositis. *Muscle and Nerve*, **2009**, 40, 19-31	3.4	143
39	Overexpression of IGF-1 in muscle attenuates disease in a mouse model of spinal and bulbar muscular atrophy. *Neuron*, **2009**, 63, 316-28	13.9	179
38	Autophagy and the ubiquitin-proteasome system: collaborators in neuroprotection. *Biochimica Et Biophysica Acta - Molecular Basis of Disease*, **2008**, 1782, 691-9	6.9	238
37	The role of autophagy in age-related neurodegeneration. *NeuroSignals*, **2008**, 16, 75-84	1.9	82
36	Guidelines for the use and interpretation of assays for monitoring autophagy in higher eukaryotes. *Autophagy*, **2008**, 4, 151-75	10.2	1920
35	HDAC6 rescues neurodegeneration and provides an essential link between autophagy and the UPS. *Nature*, **2007**, 447, 859-63	50.4	932
34	Valosin-containing protein and the pathogenesis of frontotemporal dementia associated with inclusion body myopathy. *Acta Neuropathologica*, **2007**, 114, 55-61	14.3	51
33	HDAC6 at the intersection of autophagy, the ubiquitin-proteasome system and neurodegeneration. *Autophagy*, **2007**, 3, 643-5	10.2	103
32	Safety, tolerability, and pharmacokinetics of high-dose idebenone in patients with Friedreich ataxia. *Archives of Neurology*, **2007**, 64, 803-8		68
31	TDP-43 in the ubiquitin pathology of frontotemporal dementia with VCP gene mutations. *Journal of Neuropathology and Experimental Neurology*, **2007**, 66, 152-7	3.1	256
30	Measuring Friedreich ataxia: Interrater reliability of a neurologic rating scale. *Neurology*, **2005**, 64, 1261-2	8.5	239
29	Spinal Muscular Atrophy. *Neurological Disease and Therapy*, **2005**, 209-226		
28	Hsp70 dynamics in vivo: effect of heat shock and protein aggregation. *Journal of Cell Science*, **2004**, 117, 4991-5000	5.3	66
27	A screen for drugs that protect against the cytotoxicity of polyglutamine-expanded androgen receptor. *Human Molecular Genetics*, **2004**, 13, 437-46	5.6	47
26	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, 1463-8	12.6	112
25	Valproic acid increases SMN levels in spinal muscular atrophy patient cells. *Annals of Neurology*, **2003**, 54, 647-54	9.4	253
24	Polyglutamines placed into context. *Neuron*, **2003**, 38, 681-4	13.9	114

23	Aggresomes protect cells by enhancing the degradation of toxic polyglutamine-containing protein. *Human Molecular Genetics*, **2003**, 12, 749-57	5.6	328
22	Hemolytic anemia presenting as idiopathic intracranial hypertension. *Neurology*, **2002**, 59, 960-1	6.5	7
21	Rescue of polyglutamine-mediated cytotoxicity by double-stranded RNA-mediated RNA interference. *Human Molecular Genetics*, **2002**, 11, 175-84	5.6	86
20	Repeat expansion and neurological disease **2002**, 32-54		2
19	Toxic proteins in neurodegenerative disease. *Science*, **2002**, 296, 1991-5	33.3	957
18	Transcription of intermediate filament genes is enhanced in focal cortical dysplasia. *Acta Neuropathologica*, **2001**, 102, 141-8	14.3	23
17	CREB-binding protein sequestration by expanded polyglutamine. *Human Molecular Genetics*, **2000**, 9, 2197-202	5.6	412
16	Activation of HIV-1 transcription by Tat in cells derived from the CNS: evidence for the participation of NF-kappa B--a review. *Advances in Neuroimmunology*, **1994**, 4, 291-303		19
15	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, 3971-81	6.6	37
14	The tumor suppressor protein p53 strongly alters human immunodeficiency virus type 1 replication. *Journal of Virology*, **1994**, 68, 4302-13	6.6	74
13	TAR-independent activation of HIV-1 requires the activation domain but not the RNA-binding domain of Tat. *Virology*, **1993**, 195, 780-5	3.6	22
12	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, 9617-21	11.5	89
11	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, 7355-61	6.6	58
10	TAR-independent replication of human immunodeficiency virus type 1 in glial cells. *Journal of Virology*, **1992**, 66, 7522-8	6.6	33
9	Genome-Wide Analyses Identify KIF5A as a Novel ALS Gene. *SSRN Electronic Journal*,	1	2
8	Machine learning suggests polygenic contribution to cognitive dysfunction in amyotrophic lateral sclerosis		1
7	High fidelity reconstitution of stress granules and nucleoli in mammalian cellular lysate		1
6	OptoGranules reveal the evolution of stress granules to ALS-FTD pathology		3

5	Dominant toxicity of ALS/FTD-associated CHCHD10S59L is mediated by TDP-43 and PINK1	1
4	Drosophilaand Mouse Models of Hereditary Myopathy Caused by Mutations in VCP/p97230-239	
3	Ubiquitination is essential for recovery of cellular activities following heat shock	3
2	Ubiquitination of G3BP1 mediates stress granule disassembly in a stress-specific manner	1
1	Specific heterozygous frameshift variants in hnRNPA2B1 cause early-onset oculopharyngeal muscular dystrophy	1