David R Borchelt

List of Publications by Citations

188 papers	21,869 citations	67 h-index	147 g-index
198 ext. papers	23,642 ext. citations	8.4 avg, IF	6.19 L-index

#	Paper	IF	Citations
188	Familial Alzheimer's disease-linked presenilin 1 variants elevate Abeta1-42/1-40 ratio in vitro and in vivo. *Neuron*, **1996**, 17, 1005-13	13.9	1350
187	An adverse property of a familial ALS-linked SOD1 mutation causes motor neuron disease characterized by vacuolar degeneration of mitochondria. *Neuron*, **1995**, 14, 1105-16	13.9	1281
186	APP processing and synaptic function. *Neuron*, **2003**, 37, 925-37	13.9	1248
185	Mutant presenilins specifically elevate the levels of the 42 residue beta-amyloid peptide in vivo: evidence for augmentation of a 42-specific gamma secretase. *Human Molecular Genetics*, **2004**, 13, 159-70	5.6	1073
184	Endoproteolysis of presenilin 1 and accumulation of processed derivatives in vivo. *Neuron*, **1996**, 17, 181-90	13.9	999
183	BACE1 is the major beta-secretase for generation of Abeta peptides by neurons. *Nature Neuroscience*, **2001**, 4, 233-4	25.5	935
182	Accelerated amyloid deposition in the brains of transgenic mice coexpressing mutant presenilin 1 and amyloid precursor proteins. *Neuron*, **1997**, 19, 939-45	13.9	885
181	Co-expression of multiple transgenes in mouse CNS: a comparison of strategies. *New Biotechnology* , **2001**, 17, 157-65		594
180	Decreased expression of striatal signaling genes in a mouse model of Huntington's disease. *Human Molecular Genetics*, **2000**, 9, 1259-71	5.6	550
179	Age-related CNS disorder and early death in transgenic FVB/N mice overexpressing Alzheimer amyloid precursor proteins. *Neuron*, **1995**, 15, 1203-18	13.9	482
178	BACE1, a major determinant of selective vulnerability of the brain to amyloid-beta amyloidogenesis, is essential for cognitive, emotional, and synaptic functions. *Journal of Neuroscience*, **2005**, 25, 11693-709	6.6	436
177	Environmental enrichment mitigates cognitive deficits in a mouse model of Alzheimer's disease. *Journal of Neuroscience*, **2005**, 25, 5217-24	6.6	386
176	Protein topology of presenilin 1. *Neuron*, **1996**, 17, 1023-30	13.9	358
175	Alzheimer's disease: genetic studies and transgenic models. *Annual Review of Genetics*, **1998**, 32, 461-93	14.5	349
174	Effects of PS1 deficiency on membrane protein trafficking in neurons. *Neuron*, **1998**, 21, 1213-21	13.9	342
173	SOD1 rescues cerebral endothelial dysfunction in mice overexpressing amyloid precursor protein. *Nature Neuroscience*, **1999**, 2, 157-61	25.5	337
172	Episodic-like memory deficits in the APPswe/PS1dE9 mouse model of Alzheimer's disease: relationships to beta-amyloid deposition and neurotransmitter abnormalities. *Neurobiology of Disease*, **2005**, 18, 602-17	7.5	309

171	Lipopolysaccharide-induced-neuroinflammation increases intracellular accumulation of amyloid precursor protein and amyloid beta peptide in APPswe transgenic mice. *Neurobiology of Disease*, **2003**, 14, 133-45	7.5	297
170	A vector for expressing foreign genes in the brains and hearts of transgenic mice. *Genetic Analysis, Techniques and Applications*, **1996**, 13, 159-63		290
169	Evidence that levels of presenilins (PS1 and PS2) are coordinately regulated by competition for limiting cellular factors. *Journal of Biological Chemistry*, **1997**, 272, 28415-22	5.4	275
168	Copper-binding-site-null SOD1 causes ALS in transgenic mice: aggregates of non-native SOD1 delineate a common feature. *Human Molecular Genetics*, **2003**, 12, 2753-64	5.6	253
167	Creatine increase survival and delays motor symptoms in a transgenic animal model of Huntington's disease. *Neurobiology of Disease*, **2001**, 8, 479-91	7.5	237
166	Intramuscular injection of α-synuclein induces CNS α-synuclein pathology and a rapid-onset motor phenotype in transgenic mice. *Proceedings of the National Academy of Sciences of the United States of America*, **2014**, 111, 10732-7	11.5	229
165	C9orf72 BAC Mouse Model with Motor Deficits and Neurodegenerative Features of ALS/FTD. *Neuron*, **2016**, 90, 521-34	13.9	218
164	Asparagine-linked glycosylation of the scrapie and cellular prion proteins. *Archives of Biochemistry and Biophysics*, **1989**, 274, 1-13	4.1	213
163	Fibrillar inclusions and motor neuron degeneration in transgenic mice expressing superoxide dismutase 1 with a disrupted copper-binding site. *Neurobiology of Disease*, **2002**, 10, 128-38	7.5	207
162	RAN Translation in Huntington Disease. *Neuron*, **2015**, 88, 667-77	13.9	205
161	Genetic neurodegenerative diseases: the human illness and transgenic models. *Science*, **1998**, 282, 1079-83	33.3	196
160	Genetically engineered mouse models of neurodegenerative diseases. *Nature Neuroscience*, **2002**, 5, 633-9	25.5	190
159	Alzheimer's-type amyloidosis in transgenic mice impairs survival of newborn neurons derived from adult hippocampal neurogenesis. *Journal of Neuroscience*, **2007**, 27, 6771-80	6.6	185
158	Endoproteolytic processing and stabilization of wild-type and mutant presenilin. *Journal of Biological Chemistry*, **1997**, 272, 24536-41	5.4	175
157	High molecular weight complexes of mutant superoxide dismutase 1: age-dependent and tissue-specific accumulation. *Neurobiology of Disease*, **2002**, 9, 139-48	7.5	175
156	Stable association of presenilin derivatives and absence of presenilin interactions with APP. *Neurobiology of Disease*, **1998**, 4, 438-53	7.5	175
155	Variation in aggregation propensities among ALS-associated variants of SOD1: correlation to human disease. *Human Molecular Genetics*, **2009**, 18, 3217-26	5.6	173
154	Environmental enrichment exacerbates amyloid plaque formation in a transgenic mouse model of Alzheimer disease. *Journal of Neuropathology and Experimental Neurology*, **2003**, 62, 1220-7	3.1	165

153	Polyglutamine and transcription: gene expression changes shared by DRPLA and Huntington's disease mouse models reveal context-independent effects. *Human Molecular Genetics*, **2002**, 11, 1927-37	5.6	160
152	Nuclear accumulation of truncated atrophin-1 fragments in a transgenic mouse model of DRPLA. *Neuron*, **1999**, 24, 275-86	13.9	158
151	Persistent amyloidosis following suppression of Abeta production in a transgenic model of Alzheimer disease. *PLoS Medicine*, **2005**, 2, e355	11.6	157
150	Amyloid precursor proteins inhibit heme oxygenase activity and augment neurotoxicity in Alzheimer's disease. *Neuron*, **2000**, 28, 461-73	13.9	152
149	Role of mutant SOD1 disulfide oxidation and aggregation in the pathogenesis of familial ALS. *Proceedings of the National Academy of Sciences of the United States of America*, **2009**, 106, 7774-9	11.5	149
148	Coenzyme Q10 and remacemide hydrochloride ameliorate motor deficits in a Huntington's disease transgenic mouse model. *Neuroscience Letters*, **2001**, 315, 149-53	3.3	143
147	Messenger RNA oxidation occurs early in disease pathogenesis and promotes motor neuron degeneration in ALS. *PLoS ONE*, **2008**, 3, e2849	3.7	143
146	Amyloid pathology is associated with progressive monoaminergic neurodegeneration in a transgenic mouse model of Alzheimer's disease. *Journal of Neuroscience*, **2008**, 28, 13805-14	6.6	140
145	Early phenotypes that presage late-onset neurodegenerative disease allow testing of modifiers in Hdh CAG knock-in mice. *Human Molecular Genetics*, **2002**, 11, 633-40	5.6	134
144	Hyperaccumulation of FAD-linked presenilin 1 variants in vivo. *Nature Medicine*, **1997**, 3, 756-60	50.5	131
143	An Alzheimer's disease-linked PS1 variant rescues the developmental abnormalities of PS1-deficient embryos. *Neuron*, **1998**, 20, 603-9	13.9	128
142	Superoxide dismutase 1 subunits with mutations linked to familial amyotrophic lateral sclerosis do not affect wild-type subunit function. *Journal of Biological Chemistry*, **1995**, 270, 3234-8	5.4	124
141	APP processing and amyloid deposition in mice haplo-insufficient for presenilin 1. *Neurobiology of Aging*, **2004**, 25, 885-92	5.6	119
140	Release of the cellular prion protein from cultured cells after loss of its glycoinositol phospholipid anchor. *Glycobiology*, **1993**, 3, 319-29	5.8	117
139	Immunoreactivity of the phosphorylated axonal neurofilament H subunit (pNF-H) in blood of ALS model rodents and ALS patients: evaluation of blood pNF-H as a potential ALS biomarker. *Journal of Neurochemistry*, **2009**, 111, 1182-91	6	109
138	Somatodendritic accumulation of misfolded SOD1-L126Z in motor neurons mediates degeneration: alphaB-crystallin modulates aggregation. *Human Molecular Genetics*, **2005**, 14, 2335-47	5.6	105
137	Capsid serotype and timing of injection determines AAV transduction in the neonatal mice brain. *PLoS ONE*, **2013**, 8, e67680	3.7	103
136	Synaptic transmission and hippocampal long-term potentiation in transgenic mice expressing FAD-linked presenilin 1. *Neurobiology of Disease*, **1999**, 6, 56-62	7.5	102

135	Copper and zinc metallation status of copper-zinc superoxide dismutase from amyotrophic lateral sclerosis transgenic mice. *Journal of Biological Chemistry*, **2011**, 286, 2795-806	5.4	98
134	Cyclooxygenase (COX)-2 and cell cycle activity in a transgenic mouse model of Alzheimer disease neuropathology. *Neurobiology of Aging*, **2002**, 23, 327-34	5.6	97
133	Prion-like propagation of mutant SOD1 misfolding and motor neuron disease spread along neuroanatomical pathways. *Acta Neuropathologica*, **2016**, 131, 103-14	14.3	96
132	Abnormal SDS-PAGE migration of cytosolic proteins can identify domains and mechanisms that control surfactant binding. *Protein Science*, **2012**, 21, 1197-209	6.3	93
131	Axonal transport of mutant superoxide dismutase 1 and focal axonal abnormalities in the proximal axons of transgenic mice. *Neurobiology of Disease*, **1998**, 5, 27-35	7.5	92
130	Coincident thresholds of mutant protein for paralytic disease and protein aggregation caused by restrictively expressed superoxide dismutase cDNA. *Neurobiology of Disease*, **2005**, 20, 943-52	7.5	90
129	Rodent A beta modulates the solubility and distribution of amyloid deposits in transgenic mice. *Journal of Biological Chemistry*, **2007**, 282, 22707-20	5.4	86
128	Nuclear-targeting of mutant huntingtin fragments produces Huntington disease-like phenotypes in transgenic mice. *Human Molecular Genetics*, **2004**, 13, 1599-610	5.6	85
127	A limited role for disulfide cross-linking in the aggregation of mutant SOD1 linked to familial amyotrophic lateral sclerosis. *Journal of Biological Chemistry*, **2008**, 283, 13528-37	5.4	84
126	Experimental transmissibility of mutant SOD1 motor neuron disease. *Acta Neuropathologica*, **2014**, 128, 791-803	14.3	80
125	Thinking laterally about neurodegenerative proteinopathies. *Journal of Clinical Investigation*, **2013**, 123, 1847-55	15.9	80
124	Immature copper-zinc superoxide dismutase and familial amyotrophic lateral sclerosis. *Experimental Biology and Medicine*, **2009**, 234, 1140-54	3.7	74
123	Dichloroacetate exerts therapeutic effects in transgenic mouse models of Huntington disease. *Annals of Neurology*, **2001**, 50, 112-7	9.4	72
122	Mapping superoxide dismutase 1 domains of non-native interaction: roles of intra- and intermolecular disulfide bonding in aggregation. *Journal of Neurochemistry*, **2006**, 96, 1277-88	6	70
121	Normal cognitive behavior in two distinct congenic lines of transgenic mice hyperexpressing mutant APP SWE. *Neurobiology of Disease*, **2003**, 12, 194-211	7.5	66
120	Detergent-insoluble aggregates associated with amyotrophic lateral sclerosis in transgenic mice contain primarily full-length, unmodified superoxide dismutase-1. *Journal of Biological Chemistry*, **2008**, 283, 8340-50	5.4	64
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118	Normal cognition in transgenic BRI2-A mice. *Molecular Neurodegeneration*, **2013**, 8, 15	19	57

117	Synphilin-1 attenuates neuronal degeneration in the A53T alpha-synuclein transgenic mouse model. *Human Molecular Genetics*, **2010**, 19, 2087-98	5.6	56
116	N-terminal proteolysis of full-length mutant huntingtin in an inducible PC12 cell model of Huntington's disease. *Cell Cycle*, **2007**, 6, 2970-81	4.7	56
115	Characterization of huntingtin pathologic fragments in human Huntington disease, transgenic mice, and cell models. *Journal of Neuropathology and Experimental Neurology*, **2007**, 66, 313-20	3.1	56
114	An examination of wild-type SOD1 in modulating the toxicity and aggregation of ALS-associated mutant SOD1. *Human Molecular Genetics*, **2010**, 19, 4774-89	5.6	55
113	Enhanced synaptic potentiation in transgenic mice expressing presenilin 1 familial Alzheimer's disease mutation is normalized with a benzodiazepine. *Neurobiology of Disease*, **2000**, 7, 54-63	7.5	53
112	Progressive phenotype and nuclear accumulation of an amino-terminal cleavage fragment in a transgenic mouse model with inducible expression of full-length mutant huntingtin. *Neurobiology of Disease*, **2006**, 21, 381-91	7.5	51
111	Behavioral abnormalities in APPSwe/PS1dE9 mouse model of AD-like pathology: comparative analysis across multiple behavioral domains. *Neurobiology of Aging*, **2015**, 36, 2519-32	5.6	46
110	Distinct behavioral and neuropathological abnormalities in transgenic mouse models of HD and DRPLA. *Neurobiology of Disease*, **2001**, 8, 405-18	7.5	46
109	Prion-like Spreading in Tauopathies. *Biological Psychiatry*, **2018**, 83, 337-346	7.9	44
108	The value of transgenic models for the study of neurodegenerative diseases. *Annals of the New York Academy of Sciences*, **2000**, 920, 179-91	6.5	44
107	Transgenic mouse models of neurodegenerative disease: opportunities for therapeutic development. *Current Neurology and Neuroscience Reports*, **2002**, 2, 457-64	6.6	44
106	Therapeutic approaches targeting Apolipoprotein E function in Alzheimer's disease. *Molecular Neurodegeneration*, **2020**, 15, 8	19	43
105	Papillomavirus-like particles are an effective platform for amyloid-beta immunization in rabbits and transgenic mice. *Journal of Immunology*, **2006**, 177, 2662-2670	5.3	43
104	Superoxide dismutase 1 encoding mutations linked to ALS adopts a spectrum of misfolded states. *Molecular Neurodegeneration*, **2011**, 6, 77	19	42
103	Metal-deficient aggregates and diminished copper found in cells expressing SOD1 mutations that cause ALS. *Frontiers in Aging Neuroscience*, **2014**, 6, 110	5.3	41
102	Modulation of mutant superoxide dismutase 1 aggregation by co-expression of wild-type enzyme. *Journal of Neurochemistry*, **2009**, 108, 1009-18	6	41
101	Differential regulation of small heat shock proteins in transgenic mouse models of neurodegenerative diseases. *Neurobiology of Aging*, **2008**, 29, 586-97	5.6	41
100	Disease-associated mutations at copper ligand histidine residues of superoxide dismutase 1 diminish the binding of copper and compromise dimer stability. *Journal of Biological Chemistry*, **2007**, 282, 345-52	5.4	41

99	beta-Amyloid peptide vaccination results in marked changes in serum and brain Abeta levels in APPswe/PS1DeltaE9 mice, as detected by SELDI-TOF-based ProteinChip technology. *DNA and Cell Biology*, **2001**, 20, 713-21	3.6	40
98	Reversible pathologic and cognitive phenotypes in an inducible model of Alzheimer-amyloidosis. *Journal of Neuroscience*, **2013**, 33, 3765-79	6.6	39
97	Amyloid precursor protein increases cortical neuron size in transgenic mice. *Neurobiology of Aging*, **2009**, 30, 1238-44	5.6	38
96	Widespread and efficient transduction of spinal cord and brain following neonatal AAV injection and potential disease modifying effect in ALS mice. *Molecular Therapy*, **2015**, 23, 53-62	11.7	35
95	Conformational specificity of the C4F6 SOD1 antibody; low frequency of reactivity in sporadic ALS cases. *Acta Neuropathologica Communications*, **2014**, 2, 55	7.3	35
94	Subcellular Localization of Matrin 3 Containing Mutations Associated with ALS and Distal Myopathy. *PLoS ONE*, **2015**, 10, e0142144	3.7	35
93	Structural similarity of wild-type and ALS-mutant superoxide dismutase-1 fibrils using limited proteolysis and atomic force microscopy. *Proceedings of the National Academy of Sciences of the United States of America*, **2013**, 110, 10934-9	11.5	35
92	Transgenic mice expressing caspase-6-derived N-terminal fragments of mutant huntingtin develop neurologic abnormalities with predominant cytoplasmic inclusion pathology composed largely of a smaller proteolytic derivative. *Human Molecular Genetics*, **2011**, 20, 2770-82	5.6	35
91	Cytosolic proteins lose solubility as amyloid deposits in a transgenic mouse model of Alzheimer-type amyloidosis. *Human Molecular Genetics*, **2013**, 22, 2765-74	5.6	34
90	Murine A□ over-production produces diffuse and compact Alzheimer-type amyloid deposits. *Acta Neuropathologica Communications*, **2015**, 3, 72	7.3	34
89	Motor neuron disease caused by mutations in superoxide dismutase 1. *Current Opinion in Neurology* , **1995**, 8, 294-301	7.1	33
88	Short A□ peptides attenuate A□42 toxicity in vivo. *Journal of Experimental Medicine*, **2018**, 215, 283-301	16.6	33
87	Distinct conformers of transmissible misfolded SOD1 distinguish human SOD1-FALS from other forms of familial and sporadic ALS. *Acta Neuropathologica*, **2016**, 132, 827-840	14.3	32
86	Cellular fusion for gene delivery to SCA1 affected Purkinje neurons. *Molecular and Cellular Neurosciences*, **2011**, 47, 61-70	4.8	31
85	Characterization of Protein Structural Changes in Living Cells Using Time-Lapsed FTIR Imaging. *Analytical Chemistry*, **2015**, 87, 6025-31	7.8	30
84	Rapid detection of protein aggregates in the brains of Alzheimer patients and transgenic mouse models of amyloidosis. *Alzheimer Disease and Associated Disorders*, **2002**, 16, 191-5	2.5	30
83	Unbiased screen reveals ubiquilin-1 and -2 highly associated with huntingtin inclusions. *Brain Research*, **2013**, 1524, 62-73	3.7	29
82	Analysis of chaperone mRNA expression in the adult mouse brain by meta analysis of the Allen Brain Atlas. *PLoS ONE*, **2010**, 5, e13675	3.7	26

81	Transgenic models of neurodegenerative diseases. *Current Opinion in Neurobiology*, **1996**, 6, 651-60	7.6	26
80	Characterization of gene regulation and protein interaction networks for Matrin 3 encoding mutations linked to amyotrophic lateral sclerosis and myopathy. *Scientific Reports*, **2018**, 8, 4049	4.9	25
79	Regenerative medicine in Alzheimer's disease. *Translational Research*, **2014**, 163, 432-8	11	25
78	Identification of proteins sensitive to thermal stress in human neuroblastoma and glioma cell lines. *PLoS ONE*, **2012**, 7, e49021	3.7	25
77	Quantitative Comparison of Dense-Core Amyloid Plaque Accumulation in Amyloid-β Protein Precursor Transgenic Mice. *Journal of Alzheimer's Disease*, **2017**, 56, 743-761	4.3	22
76	Amyotrophic lateral sclerosis--are microglia killing motor neurons?. *New England Journal of Medicine*, **2006**, 355, 1611-3	59.2	22
75	Role of disulfide cross-linking of mutant SOD1 in the formation of inclusion-body-like structures. *PLoS ONE*, **2012**, 7, e47838	3.7	21
74	Robust cytoplasmic accumulation of phosphorylated TDP-43 in transgenic models of tauopathy. *Acta Neuropathologica*, **2013**, 126, 39-50	14.3	20
73	Partial depletion of CREB-binding protein reduces life expectancy in a mouse model of Huntington disease. *Journal of Neuropathology and Experimental Neurology*, **2010**, 69, 396-404	3.1	20
72	Transgenic mouse models of Alzheimer's disease and amyotrophic lateral sclerosis. *Brain Pathology*, **1998**, 8, 735-57	6	20
71	Comparative analyses of the in vivo induction and transmission of α-synuclein pathology in transgenic mice by MSA brain lysate and recombinant α-synuclein fibrils. *Acta Neuropathologica Communications*, **2019**, 7, 80	7.3	19
70	N-terminal sequences in matrin 3 mediate phase separation into droplet-like structures that recruit TDP43 variants lacking RNA binding elements. *Laboratory Investigation*, **2019**, 99, 1030-1040	5.9	19
69	Changes in proteome solubility indicate widespread proteostatic disruption in mouse models of neurodegenerative disease. *Acta Neuropathologica*, **2018**, 136, 919-938	14.3	19
68	PMP22 Regulates Cholesterol Trafficking and ABCA1-Mediated Cholesterol Efflux. *Journal of Neuroscience*, **2019**, 39, 5404-5418	6.6	18
67	An examination of alpha B-crystallin as a modifier of SOD1 aggregate pathology and toxicity in models of familial amyotrophic lateral sclerosis. *Journal of Neurochemistry*, **2010**, 113, 1092-100	6	18
66	Aggregation modulating elements in mutant human superoxide dismutase 1. *Archives of Biochemistry and Biophysics*, **2010**, 503, 175-82	4.1	18
65	Premature death and neurologic abnormalities in transgenic mice expressing a mutant huntingtin exon-2 fragment. *Human Molecular Genetics*, **2011**, 20, 1633-42	5.6	18
64	Vulnerability of newly synthesized proteins to proteostasis stress. *Journal of Cell Science*, **2016**, 129, 1892-901	5.3	18

63	Non-prion-type transmission in A53T -synuclein transgenic mice: a normal component of spinal homogenates from naïve non-transgenic mice induces robust -synuclein pathology. *Acta Neuropathologica*, **2016**, 131, 151-4	14.3	17
62	A novel variant of human superoxide dismutase 1 harboring amyotrophic lateral sclerosis-associated and experimental mutations in metal-binding residues and free cysteines lacks toxicity in vivo. *Journal of Neurochemistry*, **2012**, 121, 475-85	6	17
61	A preclinical assessment of neural stem cells as delivery vehicles for anti-amyloid therapeutics. *PLoS ONE*, **2012**, 7, e34097	3.7	17
60	Limited clearance of pre-existing amyloid plaques after intracerebral injection of Abeta antibodies in two mouse models of Alzheimer disease. *Journal of Neuropathology and Experimental Neurology*, **2008**, 67, 30-40	3.1	17
59	Distinctive features of the D101N and D101G variants of superoxide dismutase 1; two mutations that produce rapidly progressing motor neuron disease. *Journal of Neurochemistry*, **2014**, 128, 305-14	6	16
58	Abeta deposition does not cause the aggregation of endogenous tau in transgenic mice. *Alzheimer Disease and Associated Disorders*, **2002**, 16, 196-201	2.5	16
57	Accumulation of proteolytic fragments of mutant presenilin 1 and accelerated amyloid deposition are co-regulated in transgenic mice. *Neurobiology of Aging*, **2002**, 23, 171-7	5.6	16
56	Reduction of low-density lipoprotein receptor-related protein (LRP1) in hippocampal neurons does not proportionately reduce, or otherwise alter, amyloid deposition in APPswe/PS1dE9 transgenic mice. *Alzheimerls Research and Therapy*, **2012**, 4, 12	9	15
55	Analysis of spinal and muscle pathology in transgenic mice overexpressing wild-type and ALS-linked mutant MATR3. *Acta Neuropathologica Communications*, **2018**, 6, 137	7.3	15
54	Aberrant accrual of BIN1 near Alzheimer disease amyloid deposits in transgenic models. *Brain Pathology*, **2019**, 29, 485-501	6	13
53	Direct and indirect mechanisms for wild-type SOD1 to enhance the toxicity of mutant SOD1 in bigenic transgenic mice. *Human Molecular Genetics*, **2015**, 24, 1019-35	5.6	12
52	Passive (amyloid-)immunotherapy attenuates monoaminergic axonal degeneration in the APPswe/PS1dE9 mice. *Journal of Alzheimerls Disease*, **2011**, 23, 271-9	4.3	12
51	Loss of functional prion protein: a role in prion disorders?. *Chemistry and Biology*, **1996**, 3, 619-21		12
50	ALS-Linked SOD1 Mutants Enhance Neurite Outgrowth and Branching in Adult Motor Neurons. *IScience*, **2019**, 11, 294-304	6.1	12
49	Reactive astrocytes as treatment targets in Alzheimer disease-Systematic review of studies using the APPswePS1dE9 mouse model. *Glia*, **2021**, 69, 1852-1881	9	12
48	Features of wild-type human SOD1 limit interactions with misfolded aggregates of mouse G86R Sod1. *Molecular Neurodegeneration*, **2013**, 8, 46	19	11
47	Relationship between mutant Cu/Zn superoxide dismutase 1 maturation and inclusion formation in cell models. *Journal of Neurochemistry*, **2017**, 140, 140-150	6	11
46	Genetically engineered models relevant to neurodegenerative disorders: their value for understanding disease mechanisms and designing/testing experimental therapeutics. *Journal of Molecular Neuroscience*, **2001**, 17, 233-57	3.3	11

45	Heterogeneity of Matrin 3 in the developing and aging murine central nervous system. *Journal of Comparative Neurology*, **2016**, 524, 2740-52	3.4	11
44	Substantially elevating the levels of B-crystallin in spinal motor neurons of mutant SOD1 mice does not significantly delay paralysis or attenuate mutant protein aggregation. *Journal of Neurochemistry*, **2015**, 133, 452-64	6	10
43	Analysis of proteolytic processes and enzymatic activities in the generation of huntingtin n-terminal fragments in an HEK293 cell model. *PLoS ONE*, **2012**, 7, e50750	3.7	10
42	Sex-related dimorphism in dentate gyrus atrophy and behavioral phenotypes in an inducible tTa:APPsi transgenic model of Alzheimer's disease. *Neurobiology of Disease*, **2016**, 96, 171-185	7.5	10
41	Tryptophan residue 32 in human Cu-Zn superoxide dismutase modulates prion-like propagation and strain selection. *PLoS ONE*, **2020**, 15, e0227655	3.7	9
40	Transgenic mice overexpressing the ALS-linked protein Matrin 3 develop a profound muscle phenotype. *Acta Neuropathologica Communications*, **2016**, 4, 122	7.3	9
39	Receptor-associated protein (RAP) plays a central role in modulating Abeta deposition in APP/PS1 transgenic mice. *PLoS ONE*, **2008**, 3, e3159	3.7	9
38	Differences in memory development among C57BL/6NCrl, 129S2/SvPasCrl, and FVB/NCrl mice after delay and trace fear conditioning. *Comparative Medicine*, **2014**, 64, 4-12	1.6	9
37	Inherited neurodegenerative diseases and transgenic models. *Brain Pathology*, **1996**, 6, 467-80	6	8
36	An analysis of interactions between fluorescently-tagged mutant and wild-type SOD1 in intracellular inclusions. *PLoS ONE*, **2013**, 8, e83981	3.7	7
35	Abeta deposition is essential to AD neuropathology. *Journal of Alzheimerls Disease*, **2002**, 4, 133-8	4.3	6
34	Influence of base-pairing in the leader region on in vitro translation of Rous sarcoma virus RNA. *Virus Research*, **1985**, 3, 141-51	6.4	6
33	Experimental Mutations in Superoxide Dismutase 1 Provide Insight into Potential Mechanisms Involved in Aberrant Aggregation in Familial Amyotrophic Lateral Sclerosis. *G3: Genes, Genomes, Genetics*, **2019**, 9, 719-728	3.2	6
32	Diversity in A deposit morphology and secondary proteome insolubility across models of Alzheimer-type amyloidosis. *Acta Neuropathologica Communications*, **2020**, 8, 43	7.3	6
31	Analysis of mutant SOD1 electrophoretic mobility by Blue Native gel electrophoresis; evidence for soluble multimeric assemblies. *PLoS ONE*, **2014**, 9, e104583	3.7	5
30	Blood-based biomarkers of inflammation in amyotrophic lateral sclerosis.. *Molecular Neurodegeneration*, **2022**, 17, 11	19	5
29	Loss of charge mutations in solvent exposed Lys residues of superoxide dismutase 1 do not induce inclusion formation in cultured cell models. *PLoS ONE*, **2018**, 13, e0206751	3.7	5
28	Subcellular diversion of cholesterol by gain- and loss-of-function mutations in PMP22. *Glia*, **2020**, 68, 2300-2315	9	4

27	Intracerebral Expression of AAV-APOE4 Is Not Sufficient to Alter Tau Burden in Two Distinct Models of Tauopathy. *Molecular Neurobiology*, **2020**, 57, 1986-2001	6.2	4
26	Phenotypic diversity in ALS and the role of poly-conformational protein misfolding. *Acta Neuropathologica*, **2021**, 142, 41-55	14.3	4
25	Targeting the accomplice to thwart the culprit: a new target for the prevention of amyloid deposition. *Journal of Clinical Investigation*, **2018**, 128, 1734-1736	15.9	3
24	IL-10 based immunomodulation initiated at birth extends lifespan in a familial mouse model of amyotrophic lateral sclerosis. *Scientific Reports*, **2020**, 10, 20862	4.9	3
23	Collusion of -Synuclein and A aggravating co-morbidities in a novel prion-type mouse model. *Molecular Neurodegeneration*, **2021**, 16, 63	19	3
22	Copper-Zinc Superoxide Dismutase, Its Copper Chaperone, and Familial Amyotrophic Lateral Sclerosis **2010**, 381-401		2
21	Transgenic and gene-targeting approaches to model disorders of motor neurons. *Seminars in Neuroscience*, **1996**, 8, 163-169		2
20	Differential induction of mutant SOD1 misfolding and aggregation by tau and -synuclein pathology. *Molecular Neurodegeneration*, **2018**, 13, 23	19	2
19	Retraction Note: Transgenic mice overexpressing the ALS-linked protein Matrin 3 develop a profound muscle phenotype. *Acta Neuropathologica Communications*, **2017**, 5, 97	7.3	1
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17	Experimental mutagenesis of huntingtin to map cleavage sites: different outcomes in cell and mouse models. *Journal of Huntingtonls Disease*, **2014**, 3, 73-86	1.9	1
16	Gabriele Schilling (September 5, 1968-July 4, 2014). *Journal of Huntingtonls Disease*, **2014**, 3, 225-7	1.9	1
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14	Selected genetically engineered models relevant to human neurodegenerative disease **2005**, 176-195		1
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