David R Borchelt

List of Publications by Year in Descending Order

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	Blood-based biomarkers of inflammation in amyotrophic lateral sclerosis.. *Molecular Neurodegeneration*, **2022**, 17, 11	19	5
187	Impact of APOE genotype on prion-type propagation of tauopathy.. *Acta Neuropathologica Communications*, **2022**, 10, 57	7.3	0
186	Pathogenic tau recruits wild-type tau into brain inclusions and induces gut degeneration in transgenic SPAM mice.. *Communications Biology*, **2022**, 5, 446	6.7	
185	Phenotypic diversity in ALS and the role of poly-conformational protein misfolding. *Acta Neuropathologica*, **2021**, 142, 41-55	14.3	4
184	Supercharging Prions via Amyloid-Selective Lysine Acetylation. *Angewandte Chemie*, **2021**, 133, 15196-15206	3.6	
183	Variation in the vulnerability of mice expressing human superoxide dismutase 1 to prion-like seeding: a study of the influence of primary amino acid sequence. *Acta Neuropathologica Communications*, **2021**, 9, 92	7.3	0
182	Supercharging Prions via Amyloid-Selective Lysine Acetylation. *Angewandte Chemie - International Edition*, **2021**, 60, 15069-15079	16.4	0
181	Novel SOD1 monoclonal antibodies against the electrostatic loop preferentially detect misfolded SOD1 aggregates. *Neuroscience Letters*, **2021**, 742, 135553	3.3	1
180	Reactive astrocytes as treatment targets in Alzheimer's disease-Systematic review of studies using the APPswePS1dE9 mouse model. *Glia*, **2021**, 69, 1852-1881	9	12
179	Remodeling Alzheimer-amyloidosis models by seeding. *Molecular Neurodegeneration*, **2021**, 16, 8	19	0
178	Collusion of □Synuclein and A□aggravating co-morbidities in a novel prion-type mouse model. *Molecular Neurodegeneration*, **2021**, 16, 63	19	3
177	Subcellular diversion of cholesterol by gain- and loss-of-function mutations in PMP22. *Glia*, **2020**, 68, 2300-2315	9	4
176	Tryptophan residue 32 in human Cu-Zn superoxide dismutase modulates prion-like propagation and strain selection. *PLoS ONE*, **2020**, 15, e0227655	3.7	9
175	Therapeutic approaches targeting Apolipoprotein E function in Alzheimer's disease. *Molecular Neurodegeneration*, **2020**, 15, 8	19	43
174	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
173	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
172	Diversity in A□deposit morphology and secondary proteome insolubility across models of Alzheimer-type□amyloidosis. *Acta Neuropathologica Communications*, **2020**, 8, 43	7.3	6

171	Comparative analyses of the in vivo induction and transmission of □synuclein pathology in transgenic mice by MSA brain lysate and recombinant □synuclein fibrils. *Acta Neuropathologica Communications*, **2019**, 7, 80	7.3	19
170	PMP22 Regulates Cholesterol Trafficking and ABCA1-Mediated Cholesterol Efflux. *Journal of Neuroscience*, **2019**, 39, 5404-5418	6.6	18
169	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
168	ALS-Linked SOD1 Mutants Enhance Neurite Outgrowth and Branching in Adult Motor Neurons. *IScience*, **2019**, 11, 294-304	6.1	12
167	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
166	Aberrant accrual of BIN1 near Alzheimer□s disease amyloid deposits in transgenic models. *Brain Pathology*, **2019**, 29, 485-501	6	13
165	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
164	Prion-like Spreading in Tauopathies. *Biological Psychiatry*, **2018**, 83, 337-346	7.9	44
163	Changes in proteome solubility indicate widespread proteostatic disruption in mouse models of neurodegenerative disease. *Acta Neuropathologica*, **2018**, 136, 919-938	14.3	19
162	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
161	Short A□peptides attenuate A□2 toxicity in vivo. *Journal of Experimental Medicine*, **2018**, 215, 283-301	16.6	33
160	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
159	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
158	Differential induction of mutant SOD1 misfolding and aggregation by tau and □synuclein pathology. *Molecular Neurodegeneration*, **2018**, 13, 23	19	2
157	Quantitative Comparison of Dense-Core Amyloid Plaque Accumulation in Amyloid-□Protein Precursor Transgenic Mice. *Journal of Alzheimerls Disease*, **2017**, 56, 743-761	4.3	22
156	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
155	Relationship between mutant Cu/Zn superoxide dismutase 1 maturation and inclusion formation in cell models. *Journal of Neurochemistry*, **2017**, 140, 140-150	6	11
154	Generation of a new transgenic mouse model for assessment of tau gene silencing therapies. *Alzheimerls Research and Therapy*, **2016**, 8, 36	9	1

153	Transgenic mice overexpressing the ALS-linked protein Matrin 3 develop a profound muscle phenotype. *Acta Neuropathologica Communications*, **2016**, 4, 122	7.3	9
152	Prion-like propagation of mutant SOD1 misfolding and motor neuron disease spread along neuroanatomical pathways. *Acta Neuropathologica*, **2016**, 131, 103-14	14.3	96
151	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
150	Heterogeneity of Matrin 3 in the developing and aging murine central nervous system. *Journal of Comparative Neurology*, **2016**, 524, 2740-52	3.4	11
149	Vulnerability of newly synthesized proteins to proteostasis stress. *Journal of Cell Science*, **2016**, 129, 1892-901	5.3	18
148	C9orf72 BAC Mouse Model with Motor Deficits and Neurodegenerative Features of ALS/FTD. *Neuron*, **2016**, 90, 521-34	13.9	218
147	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
146	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
145	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
144	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
143	Characterization of Protein Structural Changes in Living Cells Using Time-Lapsed FTIR Imaging. *Analytical Chemistry*, **2015**, 87, 6025-31	7.8	30
142	RAN Translation in Huntington Disease. *Neuron*, **2015**, 88, 667-77	13.9	205
141	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
140	Murine A over-production produces diffuse and compact Alzheimer-type amyloid deposits. *Acta Neuropathologica Communications*, **2015**, 3, 72	7.3	34
139	Subcellular Localization of Matrin 3 Containing Mutations Associated with ALS and Distal Myopathy. *PLoS ONE*, **2015**, 10, e0142144	3.7	35
138	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
137	Regenerative medicine in Alzheimer's disease. *Translational Research*, **2014**, 163, 432-8	11	25
136	Experimental transmissibility of mutant SOD1 motor neuron disease. *Acta Neuropathologica*, **2014**, 128, 791-803	14.3	80

135	Conformational specificity of the C4F6 SOD1 antibody; low frequency of reactivity in sporadic ALS cases. *Acta Neuropathologica Communications*, **2014**, 2, 55	7.3	35
134	Intramuscular injection of □synuclein induces CNS □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
133	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
132	Analysis of mutant SOD1 electrophoretic mobility by Blue Native gel electrophoresis; evidence for soluble multimeric assemblies. *PLoS ONE*, **2014**, 9, e104583	3.7	5
131	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
130	Gabriele Schilling (September 5, 1968-July 4, 2014). *Journal of Huntingtonls Disease*, **2014**, 3, 225-7	1.9	1
129	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
128	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
127	Normal cognition in transgenic BRI2-A□mice. *Molecular Neurodegeneration*, **2013**, 8, 15	19	57
126	Robust cytoplasmic accumulation of phosphorylated TDP-43 in transgenic models of tauopathy. *Acta Neuropathologica*, **2013**, 126, 39-50	14.3	20
125	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
124	Features of wild-type human SOD1 limit interactions with misfolded aggregates of mouse G86R Sod1. *Molecular Neurodegeneration*, **2013**, 8, 46	19	11
123	Unbiased screen reveals ubiquilin-1 and -2 highly associated with huntingtin inclusions. *Brain Research*, **2013**, 1524, 62-73	3.7	29
122	Reversible pathologic and cognitive phenotypes in an inducible model of Alzheimer-amyloidosis. *Journal of Neuroscience*, **2013**, 33, 3765-79	6.6	39
121	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
120	Capsid serotype and timing of injection determines AAV transduction in the neonatal mice brain. *PLoS ONE*, **2013**, 8, e67680	3.7	103
119	An analysis of interactions between fluorescently-tagged mutant and wild-type SOD1 in intracellular inclusions. *PLoS ONE*, **2013**, 8, e83981	3.7	7
118	Thinking laterally about neurodegenerative proteinopathies. *Journal of Clinical Investigation*, **2013**, 123, 1847-55	15.9	80

117	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
116	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
115	A preclinical assessment of neural stem cells as delivery vehicles for anti-amyloid therapeutics. *PLoS ONE*, **2012**, 7, e34097	3.7	17
114	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
113	Role of disulfide cross-linking of mutant SOD1 in the formation of inclusion-body-like structures. *PLoS ONE*, **2012**, 7, e47838	3.7	21
112	Identification of proteins sensitive to thermal stress in human neuroblastoma and glioma cell lines. *PLoS ONE*, **2012**, 7, e49021	3.7	25
111	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
110	Cellular fusion for gene delivery to SCA1 affected Purkinje neurons. *Molecular and Cellular Neurosciences*, **2011**, 47, 61-70	4.8	31
109	Passive (amyloid-β) immunotherapy attenuates monoaminergic axonal degeneration in the APPswe/PS1dE9 mice. *Journal of Alzheimerls Disease*, **2011**, 23, 271-9	4.3	12
108	Superoxide dismutase 1 encoding mutations linked to ALS adopts a spectrum of misfolded states. *Molecular Neurodegeneration*, **2011**, 6, 77	19	42
107	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
106	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
105	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
104	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
103	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
102	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
101	Synphilin-1 attenuates neuronal degeneration in the A53T alpha-synuclein transgenic mouse model. *Human Molecular Genetics*, **2010**, 19, 2087-98	5.6	56
100	Aggregation modulating elements in mutant human superoxide dismutase 1. *Archives of Biochemistry and Biophysics*, **2010**, 503, 175-82	4.1	18

99	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
98	Copper□Zinc Superoxide Dismutase, Its Copper Chaperone, and Familial Amyotrophic Lateral Sclerosis **2010**, 381-401		2
97	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
96	Immature copper-zinc superoxide dismutase and familial amyotrophic lateral sclerosis. *Experimental Biology and Medicine*, **2009**, 234, 1140-54	3.7	74
95	Variation in aggregation propensities among ALS-associated variants of SOD1: correlation to human disease. *Human Molecular Genetics*, **2009**, 18, 3217-26	5.6	173
94	Modulation of mutant superoxide dismutase 1 aggregation by co-expression of wild-type enzyme. *Journal of Neurochemistry*, **2009**, 108, 1009-18	6	41
93	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
92	Amyloid precursor protein increases cortical neuron size in transgenic mice. *Neurobiology of Aging*, **2009**, 30, 1238-44	5.6	38
91	Differential regulation of small heat shock proteins in transgenic mouse models of neurodegenerative diseases. *Neurobiology of Aging*, **2008**, 29, 586-97	5.6	41
90	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
89	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
88	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
87	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
86	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
85	Messenger RNA oxidation occurs early in disease pathogenesis and promotes motor neuron degeneration in ALS. *PLoS ONE*, **2008**, 3, e2849	3.7	143
84	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
83	Alzheimer□-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
82	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

81	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
80	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
79	Investigation of RNA interference to suppress expression of full-length and fragment human huntingtin. *NeuroMolecular Medicine*, **2007**, 9, 145-155	4.6	
78	Biotinylated anti-A□antibody as a tool to diagnose pre-clinical stages of Alzheimer□s Disease (AD). *FASEB Journal*, **2007**, 21, A20	0.9	
77	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
76	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
75	Amyotrophic lateral sclerosis--are microglia killing motor neurons?. *New England Journal of Medicine*, **2006**, 355, 1611-3	59.2	22
74	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
73	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
72	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
71	Effects of CAG repeat length, HTT protein length and protein context on cerebral metabolism measured using magnetic resonance spectroscopy in transgenic mouse models of Huntington□s disease. *Journal of Neurochemistry*, **2005**, 95, 553-62	6	64
70	Selected genetically engineered models relevant to human neurodegenerative disease **2005**, 176-195		1
69	Persistent amyloidosis following suppression of Abeta production in a transgenic model of Alzheimer disease. *PLoS Medicine*, **2005**, 2, e355	11.6	157
68	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
67	Environmental enrichment mitigates cognitive deficits in a mouse model of Alzheimer□s disease. *Journal of Neuroscience*, **2005**, 25, 5217-24	6.6	386
66	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
65	Transgenic mouse models of neurodegenerative disease **2004**, 533-557		
64	Nuclear-targeting of mutant huntingtin fragments produces Huntington□s disease-like phenotypes in transgenic mice. *Human Molecular Genetics*, **2004**, 13, 1599-610	5.6	85

63	APP processing and amyloid deposition in mice haplo-insufficient for presenilin 1. *Neurobiology of Aging*, **2004**, 25, 885-92	5.6	119
62	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
61	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
60	Identifying new therapeutics for Huntington's disease. *Clinical Neuroscience Research*, **2003**, 3, 179-186		1
59	APP processing and synaptic function. *Neuron*, **2003**, 37, 925-37	13.9	1248
58	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
57	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
56	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
55	Abeta deposition is essential to AD neuropathology. *Journal of Alzheimerls Disease*, **2002**, 4, 133-8	4.3	6
54	Transgenic mouse models of neurodegenerative disease: opportunities for therapeutic development. *Current Neurology and Neuroscience Reports*, **2002**, 2, 457-64	6.6	44
53	Genetically engineered mouse models of neurodegenerative diseases. *Nature Neuroscience*, **2002**, 5, 633-9	25.5	190
52	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
51	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
50	Genetically engineered models of neurodegenerative diseases **2002**, 1841-1862		1
49	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
48	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
47	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
46	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

45	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
44	Cyclooxygenase (COX)-2 and cell cycle activity in a transgenic mouse model of Alzheimer's disease neuropathology. *Neurobiology of Aging*, **2002**, 23, 327-34	5.6	97
43	Co-expression of multiple transgenes in mouse CNS: a comparison of strategies. *New Biotechnology* , **2001**, 17, 157-65		594
42	Dichloroacetate exerts therapeutic effects in transgenic mouse models of Huntington's disease. *Annals of Neurology*, **2001**, 50, 112-7	9.4	72
41	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
40	BACE1 is the major beta-secretase for generation of Abeta peptides by neurons. *Nature Neuroscience*, **2001**, 4, 233-4	25.5	935
39	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
38	Distinct behavioral and neuropathological abnormalities in transgenic mouse models of HD and DRPLA. *Neurobiology of Disease*, **2001**, 8, 405-18	7.5	46
37	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
36	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
35	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
34	Decreased expression of striatal signaling genes in a mouse model of Huntington's disease. *Human Molecular Genetics*, **2000**, 9, 1259-71	5.6	550
33	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
32	Amyloid precursor proteins inhibit heme oxygenase activity and augment neurotoxicity in Alzheimer's disease. *Neuron*, **2000**, 28, 461-73	13.9	152
31	SOD1 rescues cerebral endothelial dysfunction in mice overexpressing amyloid precursor protein. *Nature Neuroscience*, **1999**, 2, 157-61	25.5	337
30	Nuclear accumulation of truncated atrophin-1 fragments in a transgenic mouse model of DRPLA. *Neuron*, **1999**, 24, 275-86	13.9	158
29	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
28	Transgenic mouse models of Alzheimer's disease and amyotrophic lateral sclerosis. *Brain Pathology*, **1998**, 8, 735-57	6	20

27	Effects of PS1 deficiency on membrane protein trafficking in neurons. *Neuron*, **1998**, 21, 1213-21	13.9	342
26	An Alzheimer's disease-linked PS1 variant rescues the developmental abnormalities of PS1-deficient embryos. *Neuron*, **1998**, 20, 603-9	13.9	128
25	Alzheimer's disease: genetic studies and transgenic models. *Annual Review of Genetics*, **1998**, 32, 461-93	14.5	349
24	Stable association of presenilin derivatives and absence of presenilin interactions with APP. *Neurobiology of Disease*, **1998**, 4, 438-53	7.5	175
23	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
22	Genetic neurodegenerative diseases: the human illness and transgenic models. *Science*, **1998**, 282, 1079-83	33	196
21	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
20	Endoproteolytic processing and stabilization of wild-type and mutant presenilin. *Journal of Biological Chemistry*, **1997**, 272, 24536-41	5.4	175
19	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
18	Hyperaccumulation of FAD-linked presenilin 1 variants in vivo. *Nature Medicine*, **1997**, 3, 756-60	50.5	131
17	Transgenic models of neurodegenerative diseases. *Current Opinion in Neurobiology*, **1996**, 6, 651-60	7.6	26
16	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
15	Protein topology of presenilin 1. *Neuron*, **1996**, 17, 1023-30	13.9	358
14	Endoproteolysis of presenilin 1 and accumulation of processed derivatives in vivo. *Neuron*, **1996**, 17, 181-90	13.9	999
13	Transgenic and gene-targeting approaches to model disorders of motor neurons. *Seminars in Neuroscience*, **1996**, 8, 163-169		2
12	Inherited neurodegenerative diseases and transgenic models. *Brain Pathology*, **1996**, 6, 467-80	6	8
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