Shihua Li

List of Publications by Year
in descending order

88 papers	5,022 citations	101543 36 h-index	98798 67 g-index
90 all docs	90 docs citations	90 times ranked	6095 citing authors

#	Article	IF	Citations
1	N-Terminal Mutant Huntingtin Associates with Mitochondria and Impairs Mitochondrial Trafficking. Journal of Neuroscience, 2008, 28, 2783-2792.	3.6	362
2	Expression of mutant huntingtin in mouse brain astrocytes causes age-dependent neurological symptoms. Proceedings of the National Academy of Sciences of the United States of America, 2009, 106, 22480-22485.	7.1	315
3	CRISPR/Cas9-mediated gene editing ameliorates neurotoxicity in mouse model of Huntingtonâ€™s disease. Journal of Clinical Investigation, 2017, 127, 2719-2724.	8.2	282
4	A Huntingtin Knockin Pig Model Recapitulates Features of Selective Neurodegeneration in Huntingtonâ€™s Disease. Cell, 2018, 173, 989-1002.e13.	28.9	231
5	Functional disruption of the dystrophin gene in rhesus monkey using CRISPR/Cas9. Human Molecular Genetics, 2015, 24, 3764-3774.	2.9	209
6	Differential Activities of the Ubiquitinâ€“Proteasome System in Neurons versus Glia May Account for the Preferential Accumulation of Misfolded Proteins in Neurons. Journal of Neuroscience, 2008, 28, 13285-13295.	3.6	158
7	Polyglutamine domain modulates the TBP-TFIIB interaction: implications for its normal function and neurodegeneration. Nature Neuroscience, 2007, 10, 1519-1528.	14.8	147
8	Expression of Huntington's disease protein results in apoptotic neurons in the brains of cloned transgenic pigs. Human Molecular Genetics, 2010, 19, 3983-3994.	2.9	140
9	Accumulation of N-terminal mutant huntingtin in mouse and monkey models implicated as a pathogenic mechanism in Huntington's disease. Human Molecular Genetics, 2008, 17, 2738-2751.	2.9	139
10	Mutant Huntingtin Downregulates Myelin Regulatory Factor-Mediated Myelin Gene Expression and Affects Mature Oligodendrocytes. Neuron, 2015, 85, 1212-1226.	8.1	138
11	Mutant Huntingtin in Glial Cells Exacerbates Neurological Symptoms of Huntington Disease Mice. Journal of Biological Chemistry, 2010, 285, 10653-10661.	3.4	134
12	Ablation of huntingtin in adult neurons is nondeleterious but its depletion in young mice causes acute pancreatitis. Proceedings of the National Academy of Sciences of the United States of America, 2016, 113, 3359-3364.	7.1	120
13	Promoting Cas9 degradation reduces mosaic mutations in non-human primate embryos. Scientific Reports, 2017, 7, 42081.	3.3	106
14	Differential ubiquitination and degradation of huntingtin fragments modulated by ubiquitin-protein ligase E3A. Proceedings of the National Academy of Sciences of the United States of America, 2014, 111, 5706-5711.	7.1	91
15	Age-Dependent Decrease in Chaperone Activity Impairs MANF Expression, Leading to Purkinje Cell Degeneration in Inducible SCA17 Mice. Neuron, 2014, 81, 349-365.	8.1	90
16	Multiple pathways contribute to the pathogenesis of Huntington disease. Molecular Neurodegeneration, 2006, 1, 19.	10.8	89
17	Proteasomal dysfunction in aging and Huntington disease. Neurobiology of Disease, 2011, 43, 4-8.	4.4	88
18	Mutant Huntingtin Impairs BDNF Release from Astrocytes by Disrupting Conversion of Rab3a-GTP into Rab3a-GDP. Journal of Neuroscience, 2016, 36, 8790-8801.	3.6	83

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19	Early Parkinson's disease symptoms in Â-synuclein transgenic monkeys. Human Molecular Genetics, 2015, 24, 2308-2317.	2.9	82
20	Therapeutic Effect of Berberine on Huntingtonâ€™s Disease Transgenic Mouse Model. PLoS ONE, 2015, 10, e0134142.	2.5	81
21	Inhibiting the ubiquitinâ€“proteasome system leads to preferential accumulation of toxic N-terminal mutant huntingtin fragments. Human Molecular Genetics, 2010, 19, 2445-2455.	2.9	73
22	Therapeutic potential of berberine against neurodegenerative diseases. Science China Life Sciences, 2015, 58, 564-569.	4.9	71
23	Mutant Huntingtin Inhibits Î±B-Crystallin Expression and Impairs Exosome Secretion from Astrocytes. Journal of Neuroscience, 2017, 37, 9550-9563.	3.6	68
24	Huntingtin-associated Protein-1 Interacts with Pro-brain-derived Neurotrophic Factor and Mediates Its Transport and Release. Journal of Biological Chemistry, 2010, 285, 5614-5623.	3.4	65
25	Polyglutamine Expansion Reduces the Association of TATA-binding Protein with DNA and Induces DNA Binding-independent Neurotoxicity. Journal of Biological Chemistry, 2008, 283, 8283-8290.	3.4	64
26	Impaired mitochondrial trafficking in Huntington's disease. Biochimica Et Biophysica Acta - Molecular Basis of Disease, 2010, 1802, 62-65.	3.8	59
27	Neuronal expression of TATA box-binding protein containing expanded polyglutamine in knock-in mice reduces chaperone protein response by impairing the function of nuclear factor-Y transcription factor. Brain, 2011, 134, 1943-1958.	7.6	58
28	Mutant Alpha-Synuclein Causes Age-Dependent Neuropathology in Monkey Brain. Journal of Neuroscience, 2015, 35, 8345-8358.	3.6	56
29	CRISPR/Cas9-mediated PINK1 deletion leads to neurodegeneration in rhesus monkeys. Cell Research, 2019, 29, 334-336.	12.0	55
30	Preferential accumulation of N-terminal mutant huntingtin in the nuclei of striatal neurons is regulated by phosphorylation. Human Molecular Genetics, 2011, 20, 1424-1437.	2.9	54
31	MANF regulates hypothalamic control of food intake and body weight. Nature Communications, 2017, 8, 579.	12.8	47
32	Neuronal Abelson helper integration site-1 (Ahi1) deficiency in mice alters TrkB signaling with a depressive phenotype. Proceedings of the National Academy of Sciences of the United States of America, 2010, 107, 19126-19131.	7.1	45
33	Cytoplasmic mislocalization of RNA splicing factors and aberrant neuronal gene splicing in TDP-43 transgenic pig brain. Molecular Neurodegeneration, 2015, 10, 42.	10.8	45
34	Species-dependent neuropathology in transgenic SOD1 pigs. Cell Research, 2014, 24, 464-481.	12.0	44
35	Transgenic animal models for study of the pathogenesis of Huntington’s disease and therapy. Drug Design, Development and Therapy, 2015, 9, 2179.	4.3	43
36	Sarcoglycan Alpha Mitigates Neuromuscular Junction Decline in Aged Mice by Stabilizing LRP4. Journal of Neuroscience, 2018, 38, 8860-8873.	3.6	40

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37	Large Polyglutamine Repeats Cause Muscle Degeneration in SCA17 Mice. Cell Reports, 2015, 13, 196-208.	6.4	39
38	Shortening the Half-Life of Cas9 Maintains Its Gene Editing Ability and Reduces Neuronal Toxicity. Cell Reports, 2018, 25, 2653-2659.e3.	6.4	39
39	Transcriptional dysregulation of TrkA associates with neurodegeneration in spinocerebellar ataxia type 17. Human Molecular Genetics, 2009, 18, 4141-4152.	2.9	38
40	14-3-3 Protein Interacts with Huntingtin-associated Protein 1 and Regulates Its Trafficking. Journal of Biological Chemistry, 2007, 282, 4748-4756.	3.4	37
41	Differential HspBP1 expression accounts for the greater vulnerability of neurons than astrocytes to misfolded proteins. Proceedings of the National Academy of Sciences of the United States of America, 2017, 114, E7803-E7811.	7.1	37
42	Truncation of mutant huntingtin in knock-in mice demonstrates exon1 huntingtin is a key pathogenic form. Nature Communications, 2020, 11, 2582.	12.8	37
43	Compartment-Dependent Degradation of Mutant Huntingtin Accounts for Its Preferential Accumulation in Neuronal Processes. Journal of Neuroscience, 2016, 36, 8317-8328.	3.6	36
44	Influence of Species Differences on the Neuropathology of Transgenic Huntington's Disease Animal Models. Journal of Genetics and Genomics, 2012, 39, 239-245.	3.9	34
45	Caspase-4 mediates cytoplasmic accumulation of TDP-43 in the primate brains. Acta Neuropathologica, 2019, 137, 919-937.	7.7	33
46	Activation of Gene Transcription by Heat Shock Protein 27 May Contribute to Its Neuronal Protection. Journal of Biological Chemistry, 2009, 284, 27944-27951.	3.4	32
47	PINK1 kinase dysfunction triggers neurodegeneration in the primate brain without impacting mitochondrial homeostasis. Protein and Cell, 2022, 13, 26-46.	11.0	32
48	Huntingtin-associated Protein-1 Deficiency in Orexin-producing Neurons Impairs Neuronal Process Extension and Leads to Abnormal Behavior in Mice. Journal of Biological Chemistry, 2010, 285, 15941-15949.	3.4	29
49	Synaptic mutant huntingtin inhibits synapsin-1 phosphorylation and causes neurological symptoms. Journal of Cell Biology, 2013, 202, 1123-1138.	5.2	29
50	Piperine ameliorates SCA17 neuropathology by reducing ER stress. Molecular Neurodegeneration, 2018, 13, 4.	10.8	29
51	TDP-43 causes differential pathology in neuronal versus glial cells in the mouse brain. Human Molecular Genetics, 2014, 23, 2678-2693.	2.9	28
52	A CRISPR monkey model unravels a unique function of PINK1 in primate brains. Molecular Neurodegeneration, 2019, 14, 17.	10.8	28
53	Loss of Hap1 selectively promotes striatal degeneration in Huntington disease mice. Proceedings of the National Academy of Sciences of the United States of America, 2020, 117, 20265-20273.	7.1	27
54	DYRK1A regulates Hap1â€“Dcaf7/WDR68 binding with implication for delayed growth in Down syndrome. Proceedings of the National Academy of Sciences of the United States of America, 2017, 114, E1224-E1233.	7.1	25

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55	Sex-dependent Effect of BAG1 in Ameliorating Motor Deficits of Huntington Disease Transgenic Mice. Journal of Biological Chemistry, 2008, 283, 16027-16036.	3.4	24
56	Loss of Ahi1 Affects Early Development by Impairing BM88/Cend1-Mediated Neuronal Differentiation. Journal of Neuroscience, 2013, 33, 8172-8184.	3.6	24
57	Lack of RAN-mediated toxicity in Huntingtonâ€™s disease knock-in mice. Proceedings of the National Academy of Sciences of the United States of America, 2020, 117, 4411-4417.	7.1	24
58	Ubiquitin-Activating Enzyme Activity Contributes to Differential Accumulation of Mutant Huntingtin in Brain and Peripheral Tissues. Journal of Neuroscience, 2014, 34, 8411-8422.	3.6	23
59	Cerebellum-enriched protein INPP5A contributes to selective neuropathology in mouse model of spinocerebellar ataxias type 17. Nature Communications, 2020, 11, 1101.	12.8	21
60	Aged monkey brains reveal the role of ubiquitin-conjugating enzyme UBE2N in the synaptosomal accumulation of mutant huntingtin. Human Molecular Genetics, 2015, 24, 1350-1362.	2.9	20
61	Molecular mechanisms underlying Spinocerebellar Ataxia 17 (SCA17) pathogenesis. Rare Diseases (Austin, Tex), 2016, 4, e1223580.	1.8	20
62	Large Animal Models of Huntingtonâ€™s Disease. Current Topics in Behavioral Neurosciences, 2013, 22, 149-160.	1.7	19
63	New pathogenic insights from large animal models of neurodegenerative diseases. Protein and Cell, 2022, 13, 707-720.	11.0	19
64	Synergistic Toxicity of Polyglutamine-Expanded TATA-Binding Protein in Glia and Neuronal Cells: Therapeutic Implications for Spinocerebellar Ataxia 17. Journal of Neuroscience, 2017, 37, 9101-9115.	3.6	18
65	Use of CRISPR/Cas9 to model brain diseases. Progress in Neuro-Psychopharmacology and Biological Psychiatry, 2018, 81, 488-492.	4.8	18
66	The Expanding Clinical Universe of Polyglutamine Disease. Neuroscientist, 2019, 25, 512-520.	3.5	17
67	Loss of huntingtin-associated protein 1 impairs insulin secretion from pancreatic Î²-cells. Cellular and Molecular Life Sciences, 2012, 69, 1305-1317.	5.4	15
68	Molecular Mechanisms and Therapeutics for SCA17. Neurotherapeutics, 2019, 16, 1097-1105.	4.4	15
69	Phosphorylation of myelin regulatory factor by <scp>PRKG</scp> 2 mediates demyelination in Huntington's disease. EMBO Reports, 2020, 21, e49783.	4.5	15
70	Ahi1 regulates the nuclear translocation of glucocorticoid receptor to modulate stress response. Translational Psychiatry, 2021, 11, 188.	4.8	15
71	Huntingtin-Associated Protein 1 in Mouse Hypothalamus Stabilizes Glucocorticoid Receptor in Stress Response. Frontiers in Cellular Neuroscience, 2020, 14, 125.	3.7	14
72	N-terminal Huntingtin Knock-In Mice: Implications of Removing the N-terminal Region of Huntingtin for Therapy. PLoS Genetics, 2016, 12, e1006083.	3.5	14

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73	Genetically modified large animal models for investigating neurodegenerative diseases. Cell and Bioscience, 2021, 11, 218.	4.8	14
74	Genetically modified rodent models of SCA17. Journal of Neuroscience Research, 2017, 95, 1540-1547.	2.9	12
75	MANF: A New Player in the Control of Energy Homeostasis, and Beyond. Frontiers in Physiology, 2018, 9, 1725.	2.8	12
76	Use of large animal models to investigate Huntington's diseases. Cell Regeneration, 2019, 8, 9-11.	2.6	11
77	Cytoplasmic TDP-43 impairs the activity of the ubiquitin-proteasome system. Experimental Neurology, 2021, 345, 113833.	4.1	11
78	Expression and Localization of Huntingtin-Associated Protein 1 (HAP1) in the Human Digestive System. Digestive Diseases and Sciences, 2019, 64, 1486-1492.	2.3	10
79	PRRT2 frameshift mutation reduces its mRNA stability resulting loss of function in paroxysmal kinesigenic dyskinesia. Biochemical and Biophysical Research Communications, 2020, 522, 553-559.	2.1	9
80	Accumulation of Endogenous Mutant Huntingtin in Astrocytes Exacerbates Neuropathology of Huntington Disease in Mice. Molecular Neurobiology, 2021, 58, 5112-5126.	4.0	9
81	Application of CRISPR/Cas9 System in Establishing Large Animal Models. Frontiers in Cell and Developmental Biology, 2022, 10, .	3.7	8
82	CRISPR: Established Editor of Human Embryos?. Cell Stem Cell, 2017, 21, 295-296.	11.1	6
83	Lack of association of somatic CAG repeat expansion with striatal neurodegeneration in HD knock-in animal models. Human Molecular Genetics, 2021, 30, 1497-1508.	2.9	5
84	SQSTM1-mediated clearance of cytoplasmic mutant TARDBP/TDP-43 in the monkey brain. Autophagy, 2022, 18, 1955-1968.	9.1	5
85	Brain Region- and Age-Dependent 5-Hydroxymethylcytosine Activity in the Non-Human Primate. Frontiers in Aging Neuroscience, 0, 14, .	3.4	5
86	Mitochondrial-Dependent and Independent Functions of PINK1. Frontiers in Cell and Developmental Biology, 0, 10, .	3.7	4
87	CRISPR-Based Genome-Editing Tools for Huntingtonâ€™s Disease Research and Therapy. Neuroscience Bulletin, 2022, 38, 1397-1408.	2.9	2
88	A huntingtinâ€“HAP1â€“PCM1 pathway in ciliogenesis. Expert Review of Proteomics, 2012, 9, 17-19.	3.0	1