

# Shihua Li

## List of Publications by Year in descending order

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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	PINK1 kinase dysfunction triggers neurodegeneration in the primate brain without impacting mitochondrial homeostasis. <i>Protein and Cell</i> , 2022, 13, 26-46.	11.0	32
2	New pathogenic insights from large animal models of neurodegenerative diseases. <i>Protein and Cell</i> , 2022, 13, 707-720.	11.0	19
3	SQSTM1-mediated clearance of cytoplasmic mutant TARDBP/TDP-43 in the monkey brain. <i>Autophagy</i> , 2022, 18, 1955-1968.	9.1	5
4	Application of CRISPR/Cas9 System in Establishing Large Animal Models. <i>Frontiers in Cell and Developmental Biology</i> , 2022, 10, .	3.7	8
5	CRISPR-Based Genome-Editing Tools for Huntington's Disease Research and Therapy. <i>Neuroscience Bulletin</i> , 2022, 38, 1397-1408.	2.9	2
6	Ahi1 regulates the nuclear translocation of glucocorticoid receptor to modulate stress response. <i>Translational Psychiatry</i> , 2021, 11, 188.	4.8	15
7	Lack of association of somatic CAG repeat expansion with striatal neurodegeneration in HD knock-in animal models. <i>Human Molecular Genetics</i> , 2021, 30, 1497-1508.	2.9	5
8	Accumulation of Endogenous Mutant Huntingtin in Astrocytes Exacerbates Neuropathology of Huntington Disease in Mice. <i>Molecular Neurobiology</i> , 2021, 58, 5112-5126.	4.0	9
9	Cytoplasmic TDP-43 impairs the activity of the ubiquitin-proteasome system. <i>Experimental Neurology</i> , 2021, 345, 113833.	4.1	11
10	Genetically modified large animal models for investigating neurodegenerative diseases. <i>Cell and Bioscience</i> , 2021, 11, 218.	4.8	14
11	PRRT2 frameshift mutation reduces its mRNA stability resulting loss of function in paroxysmal kinesigenic dyskinesia. <i>Biochemical and Biophysical Research Communications</i> , 2020, 522, 553-559.	2.1	9
12	Loss of Hap1 selectively promotes striatal degeneration in Huntington disease mice. <i>Proceedings of the National Academy of Sciences of the United States of America</i> , 2020, 117, 20265-20273.	7.1	27
13	Phosphorylation of myelin regulatory factor by PRKG2 mediates demyelination in Huntington's disease. <i>EMBO Reports</i> , 2020, 21, e49783.	4.5	15
14	Huntingtin-Associated Protein 1 in Mouse Hypothalamus Stabilizes Glucocorticoid Receptor in Stress Response. <i>Frontiers in Cellular Neuroscience</i> , 2020, 14, 125.	3.7	14
15	Lack of RAN-mediated toxicity in Huntington's disease knock-in mice. <i>Proceedings of the National Academy of Sciences of the United States of America</i> , 2020, 117, 4411-4417.	7.1	24
16	Cerebellum-enriched protein INPP5A contributes to selective neuropathology in mouse model of spinocerebellar ataxias type 17. <i>Nature Communications</i> , 2020, 11, 1101.	12.8	21
17	Truncation of mutant huntingtin in knock-in mice demonstrates exon1 huntingtin is a key pathogenic form. <i>Nature Communications</i> , 2020, 11, 2582.	12.8	37
18	Molecular Mechanisms and Therapeutics for SCA17. <i>Neurotherapeutics</i> , 2019, 16, 1097-1105.	4.4	15

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19	A CRISPR monkey model unravels a unique function of PINK1 in primate brains. <i>Molecular Neurodegeneration</i> , 2019, 14, 17.	10.8	28
20	Caspase-4 mediates cytoplasmic accumulation of TDP-43 in the primate brains. <i>Acta Neuropathologica</i> , 2019, 137, 919-937.	7.7	33
21	Use of large animal models to investigate Huntington's diseases. <i>Cell Regeneration</i> , 2019, 8, 9-11.	2.6	11
22	CRISPR/Cas9-mediated PINK1 deletion leads to neurodegeneration in rhesus monkeys. <i>Cell Research</i> , 2019, 29, 334-336.	12.0	55
23	Expression and Localization of Huntingtin-Associated Protein 1 (HAP1) in the Human Digestive System. <i>Digestive Diseases and Sciences</i> , 2019, 64, 1486-1492.	2.3	10
24	The Expanding Clinical Universe of Polyglutamine Disease. <i>Neuroscientist</i> , 2019, 25, 512-520.	3.5	17
25	A Huntingtin Knockin Pig Model Recapitulates Features of Selective Neurodegeneration in Huntington's Disease. <i>Cell</i> , 2018, 173, 989-1002.e13.	28.9	231
26	Use of CRISPR/Cas9 to model brain diseases. <i>Progress in Neuro-Psychopharmacology and Biological Psychiatry</i> , 2018, 81, 488-492.	4.8	18
27	MANF: A New Player in the Control of Energy Homeostasis, and Beyond. <i>Frontiers in Physiology</i> , 2018, 9, 1725.	2.8	12
28	Shortening the Half-Life of Cas9 Maintains Its Gene Editing Ability and Reduces Neuronal Toxicity. <i>Cell Reports</i> , 2018, 25, 2653-2659.e3.	6.4	39
29	Sarcoglycan Alpha Mitigates Neuromuscular Junction Decline in Aged Mice by Stabilizing LRP4. <i>Journal of Neuroscience</i> , 2018, 38, 8860-8873.	3.6	40
30	Piperine ameliorates SCA17 neuropathology by reducing ER stress. <i>Molecular Neurodegeneration</i> , 2018, 13, 4.	10.8	29
31	DYRK1A regulates Hap1's Dcaf7/WDR68 binding with implication for delayed growth in Down syndrome. <i>Proceedings of the National Academy of Sciences of the United States of America</i> , 2017, 114, E1224-E1233.	7.1	25
32	Promoting Cas9 degradation reduces mosaic mutations in non-human primate embryos. <i>Scientific Reports</i> , 2017, 7, 42081.	3.3	106
33	Synergistic Toxicity of Polyglutamine-Expanded TATA-Binding Protein in Glia and Neuronal Cells: Therapeutic Implications for Spinocerebellar Ataxia 17. <i>Journal of Neuroscience</i> , 2017, 37, 9101-9115.	3.6	18
34	Differential HspBP1 expression accounts for the greater vulnerability of neurons than astrocytes to misfolded proteins. <i>Proceedings of the National Academy of Sciences of the United States of America</i> , 2017, 114, E7803-E7811.	7.1	37
35	MANF regulates hypothalamic control of food intake and body weight. <i>Nature Communications</i> , 2017, 8, 579.	12.8	47
36	Mutant Huntingtin Inhibits $\beta$ -Crystallin Expression and Impairs Exosome Secretion from Astrocytes. <i>Journal of Neuroscience</i> , 2017, 37, 9550-9563.	3.6	68

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37	CRISPR: Established Editor of Human Embryos?. <i>Cell Stem Cell</i> , 2017, 21, 295-296.	11.1	6
38	Genetically modified rodent models of SCA17. <i>Journal of Neuroscience Research</i> , 2017, 95, 1540-1547.	2.9	12
39	CRISPR/Cas9-mediated gene editing ameliorates neurotoxicity in mouse model of Huntington's disease. <i>Journal of Clinical Investigation</i> , 2017, 127, 2719-2724.	8.2	282
40	Molecular mechanisms underlying Spinocerebellar Ataxia 17 (SCA17) pathogenesis. <i>Rare Diseases (Austin, Tex )</i> , 2016, 4, e1223580.	1.8	20
41	Compartment-Dependent Degradation of Mutant Huntingtin Accounts for Its Preferential Accumulation in Neuronal Processes. <i>Journal of Neuroscience</i> , 2016, 36, 8317-8328.	3.6	36
42	Mutant Huntingtin Impairs BDNF Release from Astrocytes by Disrupting Conversion of Rab3a-GTP into Rab3a-GDP. <i>Journal of Neuroscience</i> , 2016, 36, 8790-8801.	3.6	83
43	Ablation of huntingtin in adult neurons is nondeleterious but its depletion in young mice causes acute pancreatitis. <i>Proceedings of the National Academy of Sciences of the United States of America</i> , 2016, 113, 3359-3364.	7.1	120
44	N-terminal Huntingtin Knock-In Mice: Implications of Removing the N-terminal Region of Huntingtin for Therapy. <i>PLoS Genetics</i> , 2016, 12, e1006083.	3.5	14
45	Therapeutic Effect of Berberine on Huntington's Disease Transgenic Mouse Model. <i>PLoS ONE</i> , 2015, 10, e0134142.	2.5	81
46	Transgenic animal models for study of the pathogenesis of Huntington's disease and therapy. <i>Drug Design, Development and Therapy</i> , 2015, 9, 2179.	4.3	43
47	Mutant Huntingtin Downregulates Myelin Regulatory Factor-Mediated Myelin Gene Expression and Affects Mature Oligodendrocytes. <i>Neuron</i> , 2015, 85, 1212-1226.	8.1	138
48	Mutant Alpha-Synuclein Causes Age-Dependent Neuropathology in Monkey Brain. <i>Journal of Neuroscience</i> , 2015, 35, 8345-8358.	3.6	56
49	Early Parkinson's disease symptoms in $\alpha$ -synuclein transgenic monkeys. <i>Human Molecular Genetics</i> , 2015, 24, 2308-2317.	2.9	82
50	Therapeutic potential of berberine against neurodegenerative diseases. <i>Science China Life Sciences</i> , 2015, 58, 564-569.	4.9	71
51	Functional disruption of the dystrophin gene in rhesus monkey using CRISPR/Cas9. <i>Human Molecular Genetics</i> , 2015, 24, 3764-3774.	2.9	209
52	Large Polyglutamine Repeats Cause Muscle Degeneration in SCA17 Mice. <i>Cell Reports</i> , 2015, 13, 196-208.	6.4	39
53	Cytoplasmic mislocalization of RNA splicing factors and aberrant neuronal gene splicing in TDP-43 transgenic pig brain. <i>Molecular Neurodegeneration</i> , 2015, 10, 42.	10.8	45
54	Aged monkey brains reveal the role of ubiquitin-conjugating enzyme UBE2N in the synaptosomal accumulation of mutant huntingtin. <i>Human Molecular Genetics</i> , 2015, 24, 1350-1362.	2.9	20

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55	TDP-43 causes differential pathology in neuronal versus glial cells in the mouse brain. <i>Human Molecular Genetics</i> , 2014, 23, 2678-2693.	2.9	28
56	Species-dependent neuropathology in transgenic SOD1 pigs. <i>Cell Research</i> , 2014, 24, 464-481.	12.0	44
57	Differential ubiquitination and degradation of huntingtin fragments modulated by ubiquitin-protein ligase E3A. <i>Proceedings of the National Academy of Sciences of the United States of America</i> , 2014, 111, 5706-5711.	7.1	91
58	Age-Dependent Decrease in Chaperone Activity Impairs MANF Expression, Leading to Purkinje Cell Degeneration in Inducible SCA17 Mice. <i>Neuron</i> , 2014, 81, 349-365.	8.1	90
59	Ubiquitin-Activating Enzyme Activity Contributes to Differential Accumulation of Mutant Huntingtin in Brain and Peripheral Tissues. <i>Journal of Neuroscience</i> , 2014, 34, 8411-8422.	3.6	23
60	Loss of Ahi1 Affects Early Development by Impairing BM88/Cend1-Mediated Neuronal Differentiation. <i>Journal of Neuroscience</i> , 2013, 33, 8172-8184.	3.6	24
61	Synaptic mutant huntingtin inhibits synapsin-1 phosphorylation and causes neurological symptoms. <i>Journal of Cell Biology</i> , 2013, 202, 1123-1138.	5.2	29
62	Large Animal Models of Huntington's Disease. <i>Current Topics in Behavioral Neurosciences</i> , 2013, 22, 149-160.	1.7	19
63	A huntingtin-HAP1-PCM1 pathway in ciliogenesis. <i>Expert Review of Proteomics</i> , 2012, 9, 17-19.	3.0	1
64	Influence of Species Differences on the Neuropathology of Transgenic Huntington's Disease Animal Models. <i>Journal of Genetics and Genomics</i> , 2012, 39, 239-245.	3.9	34
65	Loss of huntingtin-associated protein 1 impairs insulin secretion from pancreatic $\beta$ -cells. <i>Cellular and Molecular Life Sciences</i> , 2012, 69, 1305-1317.	5.4	15
66	Proteasomal dysfunction in aging and Huntington disease. <i>Neurobiology of Disease</i> , 2011, 43, 4-8.	4.4	88
67	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- $\kappa$ B transcription factor. <i>Brain</i> , 2011, 134, 1943-1958.	7.6	58
68	Preferential accumulation of N-terminal mutant huntingtin in the nuclei of striatal neurons is regulated by phosphorylation. <i>Human Molecular Genetics</i> , 2011, 20, 1424-1437.	2.9	54
69	Neuronal Abelson helper integration site-1 (Ahi1) deficiency in mice alters TrkB signaling with a depressive phenotype. <i>Proceedings of the National Academy of Sciences of the United States of America</i> , 2010, 107, 19126-19131.	7.1	45
70	Inhibiting the ubiquitin-proteasome system leads to preferential accumulation of toxic N-terminal mutant huntingtin fragments. <i>Human Molecular Genetics</i> , 2010, 19, 2445-2455.	2.9	73
71	Huntingtin-associated Protein-1 Interacts with Pro-brain-derived Neurotrophic Factor and Mediates Its Transport and Release. <i>Journal of Biological Chemistry</i> , 2010, 285, 5614-5623.	3.4	65
72	Huntingtin-associated Protein-1 Deficiency in Orexin-producing Neurons Impairs Neuronal Process Extension and Leads to Abnormal Behavior in Mice. <i>Journal of Biological Chemistry</i> , 2010, 285, 15941-15949.	3.4	29

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73	Expression of Huntington's disease protein results in apoptotic neurons in the brains of cloned transgenic pigs. <i>Human Molecular Genetics</i> , 2010, 19, 3983-3994.	2.9	140
74	Mutant Huntingtin in Glial Cells Exacerbates Neurological Symptoms of Huntington Disease Mice. <i>Journal of Biological Chemistry</i> , 2010, 285, 10653-10661.	3.4	134
75	Impaired mitochondrial trafficking in Huntington's disease. <i>Biochimica Et Biophysica Acta - Molecular Basis of Disease</i> , 2010, 1802, 62-65.	3.8	59
76	Activation of Gene Transcription by Heat Shock Protein 27 May Contribute to Its Neuronal Protection. <i>Journal of Biological Chemistry</i> , 2009, 284, 27944-27951.	3.4	32
77	Expression of mutant huntingtin in mouse brain astrocytes causes age-dependent neurological symptoms. <i>Proceedings of the National Academy of Sciences of the United States of America</i> , 2009, 106, 22480-22485.	7.1	315
78	Transcriptional dysregulation of TrkA associates with neurodegeneration in spinocerebellar ataxia type 17. <i>Human Molecular Genetics</i> , 2009, 18, 4141-4152.	2.9	38
79	Polyglutamine Expansion Reduces the Association of TATA-binding Protein with DNA and Induces DNA Binding-independent Neurotoxicity. <i>Journal of Biological Chemistry</i> , 2008, 283, 8283-8290.	3.4	64
80	N-Terminal Mutant Huntingtin Associates with Mitochondria and Impairs Mitochondrial Trafficking. <i>Journal of Neuroscience</i> , 2008, 28, 2783-2792.	3.6	362
81	Sex-dependent Effect of BAG1 in Ameliorating Motor Deficits of Huntington Disease Transgenic Mice. <i>Journal of Biological Chemistry</i> , 2008, 283, 16027-16036.	3.4	24
82	Accumulation of N-terminal mutant huntingtin in mouse and monkey models implicated as a pathogenic mechanism in Huntington's disease. <i>Human Molecular Genetics</i> , 2008, 17, 2738-2751.	2.9	139
83	Differential Activities of the Ubiquitin-Proteasome System in Neurons versus Glia May Account for the Preferential Accumulation of Misfolded Proteins in Neurons. <i>Journal of Neuroscience</i> , 2008, 28, 13285-13295.	3.6	158
84	14-3-3 Protein Interacts with Huntingtin-associated Protein 1 and Regulates Its Trafficking. <i>Journal of Biological Chemistry</i> , 2007, 282, 4748-4756.	3.4	37
85	Polyglutamine domain modulates the TBP-TFIIB interaction: implications for its normal function and neurodegeneration. <i>Nature Neuroscience</i> , 2007, 10, 1519-1528.	14.8	147
86	Multiple pathways contribute to the pathogenesis of Huntington disease. <i>Molecular Neurodegeneration</i> , 2006, 1, 19.	10.8	89
87	Brain Region- and Age-Dependent 5-Hydroxymethylcytosine Activity in the Non-Human Primate. <i>Frontiers in Aging Neuroscience</i> , 0, 14, .	3.4	5
88	Mitochondrial-Dependent and Independent Functions of PINK1. <i>Frontiers in Cell and Developmental Biology</i> , 0, 10, .	3.7	4