Gillian P Bates

List of Publications by Year
in descending order

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233 papers	41,305 citations	4370 86 h-index	2323 199 g-index
242 all docs	242 docs citations	242 times ranked	21250 citing authors

#	Article	IF	Citations
1	Uninterrupted CAG repeat drives striatum-selective transcriptionopathy and nuclear pathogenesis in human Huntingtin BAC mice. Neuron, 2022, 110, 1173-1192.e7.	3.8	30
2	Development of novel bioassays to detect soluble and aggregated Huntingtin proteins on three technology platforms. Brain Communications, 2021, 3, fcaa231.	1.5	11
3	Ablation of kynurenine 3-monooxygenase rescues plasma inflammatory cytokine levels in the R6/2 mouse model of Huntingtonâ€™s disease. Scientific Reports, 2021, 11, 5484.	1.6	14
4	The heat shock response, determined by QuantiGene multiplex, is impaired in HD mouse models and not caused by HSF1 reduction. Scientific Reports, 2021, 11, 9117.	1.6	7
5	Use of high-content imaging to quantify transduction of AAV-PHP viruses in the brain following systemic delivery. Brain Communications, 2021, 3, fcab105.	1.5	7
6	Correlative light and electron microscopy suggests that mutant huntingtin dysregulates the endolysosomal pathway in presymptomatic Huntingtonâ€™s disease. Acta Neuropathologica Communications, 2021, 9, 70.	2.4	7
7	Small, Seeding-Competent Huntingtin Fibrils Are Prominent Aggregate Species in Brains of zQ175 Huntingtonâ€™s Disease Knock-in Mice. Frontiers in Neuroscience, 2021, 15, 682172.	1.4	7
8	Transglutaminase 6 Is Colocalized and Interacts with Mutant Huntingtin in Huntington Disease Rodent Animal Models. International Journal of Molecular Sciences, 2021, 22, 8914.	1.8	6
9	FAN1 controls mismatch repair complex assembly via MLH1 retention to stabilize CAG repeat expansion in Huntingtonâ€™s disease. Cell Reports, 2021, 36, 109649.	2.9	32
10	C02â€...FAN1 controls cag repeat expansion in huntingtonâ€™s disease by dual functions, MLH1 retention and nuclease activity. , 2021, , .		0
11	Silencing Srsf6 does not modulate incomplete splicing of the huntingtin gene in Huntingtonâ€™s disease models. Scientific Reports, 2020, 10, 14057.	1.6	17
12	Subcellular Localization And Formation Of Huntingtin Aggregates Correlates With Symptom Onset And Progression In A Huntingtonâ€™S Disease Model. Brain Communications, 2020, 2, fcaa066.	1.5	34
13	Expression of mutant exon 1 huntingtin fragments in human neural stem cells and neurons causes inclusion formation and mitochondrial dysfunction. FASEB Journal, 2020, 34, 8139-8154.	0.2	18
14	MEF2 impairment underlies skeletal muscle atrophy in polyglutamine disease. Acta Neuropathologica, 2020, 140, 63-80.	3.9	23
15	TBK1 phosphorylates mutant Huntingtin and suppresses its aggregation and toxicity in Huntington's disease models. EMBO Journal, 2020, 39, e104671.	3.5	34
16	Meso scale discovery-based assays for the detection of aggregated huntingtin. PLoS ONE, 2019, 14, e0213521.	1.1	31
17	Phenotype onset in Huntingtonâ€™s disease knockâ€in mice is correlated with the incomplete splicing of the mutant huntingtin gene. Journal of Neuroscience Research, 2019, 97, 1590-1605.	1.3	38
18	Genetic deletion of S6k1 does not rescue the phenotypic deficits observed in the R6/2 mouse model of Huntingtonâ€™s disease. Scientific Reports, 2019, 9, 16133.	1.6	2

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19	Extensive Expression Analysis of Htt Transcripts in Brain Regions from the zQ175 HD Mouse Model Using a QuantiGene Multiplex Assay. Scientific Reports, 2019, 9, 16137.	1.6	16
20	Inhibition of tumour necrosis factor alpha in the R6/2 mouse model of Huntingtonâ€™s disease by etanercept treatment. Scientific Reports, 2019, 9, 7202.	1.6	16
21	Live-cell super-resolution microscopy reveals a primary role for diffusion in polyglutamine-driven aggresome assembly. Journal of Biological Chemistry, 2019, 294, 257-268.	1.6	27
22	RNA Related Pathology in Huntingtonâ€™s Disease. Advances in Experimental Medicine and Biology, 2018, 1049, 85-101.	0.8	13
23	A04â€...The role of splicing factor SRSF6 in incomplete splicing of the HTT transcript. , 2018, , .		0
24	D05â€...Development and optimisation of a quantigene assay to measure HTT transcripts levels and investigate the efficiency of lowering htt in vivo. , 2018, , .		0
25	A03â€...Regulatory mechanisms of incomplete HTT MRNA splicing in huntingtonâ€™s disease. , 2018, , .		1
26	A18â€...Investigating the mechanisms of the heat shock response impairment in huntingtonâ€™s disease. , 2018, , .		0
27	A16â€...The temporal and spatial appearance of huntingtin aggregates in the brains of the ZQ175 mouse model of huntingtonâ€™s disease. , 2018, , .		0
28	B19â€...Development of in vitro models to investigate the pathogenesis of huntingtonâ€™s disease and screen for therapeutic agents. , 2018, , .		0
29	A21â€...Transcriptional dysregulation is caused by the accumulation of detergent insoluble HTT aggregates in the nucleus. , 2018, , .		0
30	Regulatory mechanisms of incomplete huntingtin mRNA splicing. Nature Communications, 2018, 9, 3955.	5.8	55
31	mHTT Seeding Activity: A Marker of Disease Progression and Neurotoxicity in Models of Huntingtonâ€™s Disease. Molecular Cell, 2018, 71, 675-688.e6.	4.5	50
32	Mouse Models of Huntingtonâ€™s Disease. Methods in Molecular Biology, 2018, 1780, 97-120.	0.4	57
33	In vivo neutralization of the protagonist role of macrophages during the chronic inflammatory stage of Huntingtonâ€™s disease. Scientific Reports, 2018, 8, 11447.	1.6	36
34	A20â€...A role for transglutaminase 6 in hd pathology. , 2018, , .		0
35	The pathogenic exon 1 HTT protein is produced by incomplete splicing in Huntingtonâ€™s disease patients. Scientific Reports, 2017, 7, 1307.	1.6	150
36	Myostatin inhibition prevents skeletal muscle pathophysiology in Huntingtonâ€™s disease mice. Scientific Reports, 2017, 7, 14275.	1.6	27

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37	HSF1-dependent and -independent regulation of the mammalian in vivo heat shock response and its impairment in Huntington's disease mouse models. Scientific Reports, 2017, 7, 12556.	1.6	27
38	Disruption to schizophrenia-associated gene Fez1 in the hippocampus of HDAC11 knockout mice. Scientific Reports, 2017, 7, 11900.	1.6	21
39	Stall in Canonical Autophagy-Lysosome Pathways Prompts Nucleophagy-Based Nuclear Breakdown in Neurodegeneration. Current Biology, 2017, 27, 3626-3642.e6.	1.8	47
40	Frequency of nuclear mutant huntingtin inclusion formation in neurons and glia is cellâ€typeâ€specific. Glia, 2017, 65, 50-61.	2.5	84
41	Correlations of Behavioral Deficits with Brain Pathology Assessed through Longitudinal MRI and Histopathology in the HdhQ150/Q150 Mouse Model of Huntingtonâ€™s Disease. PLoS ONE, 2017, 12, e0168556.	1.1	17
42	B4â€...Detection of the aberrantly spliced exon 1 â€“ intron 1 htt mRNA in HD patient post mortem brain tissue and fibroblast lines. Journal of Neurology, Neurosurgery and Psychiatry, 2016, 87, A10.2-A10.	0.9	0
43	B27â€...Abnormal bioenergetics in inclusion-containing mutant HTT exon 1 primary human neurons. Journal of Neurology, Neurosurgery and Psychiatry, 2016, 87, A18.2-A19.	0.9	0
44	B3â€...Comparison of the effect of a pure CAG repeat and mixed cagcaa repeat on the extent to which the htt gene is aberrantly spliced in knock-in mice. Journal of Neurology, Neurosurgery and Psychiatry, 2016, 87, A10.1-A10.	0.9	0
45	SIRT1 Activity Is Linked to Its Brain Region-Specific Phosphorylation and Is Impaired in Huntingtonâ€™s Disease Mice. PLoS ONE, 2016, 11, e0145425.	1.1	29
46	Embryonic Mutant Huntingtin Aggregate Formation in Mouse Models of Huntingtonâ€™s Disease. Journal of Huntington's Disease, 2016, 5, 343-346.	0.9	10
47	B6â€...Super-resolution fluorescence imaging of the seeding and polymerizatoin of the huntingtin exon 1 protein. Journal of Neurology, Neurosurgery and Psychiatry, 2016, 87, A11.1-A11.	0.9	0
48	The S/T-Rich Motif in the DNAJB6 Chaperone Delays Polyglutamine Aggregation and the Onset of Disease in a Mouse Model. Molecular Cell, 2016, 62, 272-283.	4.5	140
49	UBQLN2 Mediates Autophagy-Independent Protein Aggregate Clearance by the Proteasome. Cell, 2016, 166, 935-949.	13.5	248
50	L3â€...Systemic administration of a novel AAV variant results in widespread and efficient gene transfer in R6/2 mice. Journal of Neurology, Neurosurgery and Psychiatry, 2016, 87, A91.1-A91.	0.9	0
51	B10â€...Inclusion formation in mutant HTT exon 1 expressing human neuronal cells. Journal of Neurology, Neurosurgery and Psychiatry, 2016, 87, A12.2-A12.	0.9	0
52	B38â€...The effect of Hdac4 reduction post-weaning on hd-related phenotypes in R6/2 mice. Journal of Neurology, Neurosurgery and Psychiatry, 2016, 87, A22.2-A22.	0.9	3
53	B24â€...Assessment of immune system activation status during the course of disease in huntingtonâ€™s disease mouse model. Journal of Neurology, Neurosurgery and Psychiatry, 2016, 87, A17.2-A17.	0.9	0
54	B8â€...Ablation of p62 modulates levels of soluble and aggregated mutant huntingtin and delays end-stage disease in R6/2 mice. Journal of Neurology, Neurosurgery and Psychiatry, 2016, 87, A11.3-A12.	0.9	0

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55	Alan Walker (ed.), The New Science of Ageing, Policy Press, Bristol, UK, 2014, 344 pp., pbk Â£26.99, ISBN 13: 9781447314677.. Ageing and Society, 2015, 35, 1796-1797.	1.2	0
56	In Vivo Profiling Reveals a Competent Heat Shock Response in Adult Neurons: Implications for Neurodegenerative Disorders. PLoS ONE, 2015, 10, e0131985.	1.1	12
57	Characterisation of immune cell function in fragment and full-length Huntington's disease mouse models. Neurobiology of Disease, 2015, 73, 388-398.	2.1	50
58	HDAC4-Myogenin Axis As an Important Marker of HD-Related Skeletal Muscle Atrophy. PLoS Genetics, 2015, 11, e1005021.	1.5	56
59	Systematic interaction network filtering identifies CRMP1 as a novel suppressor of huntingtin misfolding and neurotoxicity. Genome Research, 2015, 25, 701-713.	2.4	24
60	Huntington disease. Nature Reviews Disease Primers, 2015, 1, 15005.	18.1	1,031
61	Treating the whole body in Huntington's disease. Lancet Neurology, The, 2015, 14, 1135-1142.	4.9	126
62	Characterization of Gastric Mucosa Biopsies Reveals Alterations in Huntington's Disease. PLOS Currents, 2015, 7, .	1.4	10
63	Novel Isoforms of Heat Shock Transcription Factor 1, HSF1Î³Î± and HSF1Î³Î², Regulate Chaperone Protein Gene Transcription. Journal of Biological Chemistry, 2014, 289, 19894-19906.	1.6	20
64	Dysfunction of the CNS-Heart Axis in Mouse Models of Huntington's Disease. PLoS Genetics, 2014, 10, e1004550.	1.5	83
65	A common gene expression signature in Huntingtonâ€™s disease patient brain regions. BMC Medical Genomics, 2014, 7, 60.	0.7	49
66	Contesting the dogma of an age-related heat shock response impairment: implications for cardiac-specific age-related disorders. Human Molecular Genetics, 2014, 23, 3641-3656.	1.4	33
67	HTT-lowering reverses Huntingtonâ€™s disease immune dysfunction caused by NFÎºB pathway dysregulation. Brain, 2014, 137, 819-833.	3.7	147
68	Dynamic recruitment of active proteasomes into polyglutamine initiated inclusion bodies. FEBS Letters, 2014, 588, 151-159.	1.3	44
69	The PDE1/5 Inhibitor SCH-51866 Does Not Modify Disease Progression in the R6/2 Mouse Model of Huntingtonâ€™s Disease. PLOS Currents, 2014, 6, .	1.4	4
70	Reducing Igf-1r Levels Leads To Paradoxical and Sexually Dimorphic Effects in HD Mice. PLoS ONE, 2014, 9, e105595.	1.1	8
71	The Huntington's Disease-Related Cardiomyopathy Prevents a Hypertrophic Response in the R6/2 Mouse Model. PLoS ONE, 2014, 9, e108961.	1.1	29
72	Downregulation of cannabinoid receptor 1 from neuropeptide <scp>Y</scp> interneurons in the basal ganglia of patients with Huntington's disease and mouse models. European Journal of Neuroscience, 2013, 37, 429-440.	1.2	46

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73	SUMO-2 and PIAS1 Modulate Insoluble Mutant Huntingtin Protein Accumulation. Cell Reports, 2013, 4, 362-375.	2.9	97
74	HDAC4 Reduction: A Novel Therapeutic Strategy to Target Cytoplasmic Huntingtin and Ameliorate Neurodegeneration. PLoS Biology, 2013, 11, e1001717.	2.6	143
75	Aberrantly splicedHTT,a new player in Huntingtonâ€™s disease pathogenesis. RNA Biology, 2013, 10, 1647-1652.	1.5	50
76	Targeting H3K4 trimethylation in Huntington disease. Proceedings of the National Academy of Sciences of the United States of America, 2013, 110, E3027-36.	3.3	151
77	Aberrant splicing of HTT generates the pathogenic exon 1 protein in Huntington disease. Proceedings of the National Academy of Sciences of the United States of America, 2013, 110, 2366-2370.	3.3	415
78	Correlations of Behavioral Deficits with Brain Pathology Assessed through Longitudinal MRI and Histopathology in the R6/2 Mouse Model of HD. PLoS ONE, 2013, 8, e60012.	1.1	44
79	HDAC4 Does Not Act as a Protein Deacetylase in the Postnatal Murine Brain In Vivo. PLoS ONE, 2013, 8, e80849.	1.1	30
80	Correlations of Behavioral Deficits with Brain Pathology Assessed through Longitudinal MRI and Histopathology in the R6/1 Mouse Model of Huntingtonâ€™s Disease. PLoS ONE, 2013, 8, e84726.	1.1	39
81	Genetic Knock-Down of Hdac3 Does Not Modify Disease-Related Phenotypes in a Mouse Model of Huntington's Disease. PLoS ONE, 2012, 7, e31080.	1.1	51
82	Suppression of protein aggregation by chaperone modification of high molecular weight complexes. Brain, 2012, 135, 1180-1196.	3.7	103
83	Implantation of undifferentiated and pre-differentiated human neural stem cells in the R6/2 transgenic mouse model of Huntingtonâ€™s disease. BMC Neuroscience, 2012, 13, 97.	0.8	40
84	TR-FRET-Based Duplex Immunoassay Reveals an Inverse Correlation of Soluble and Aggregated Mutant huntingtin in Huntington's Disease. Chemistry and Biology, 2012, 19, 264-275.	6.2	70
85	Mutant huntingtin fragmentation in immune cells tracks Huntingtonâ€™s disease progression. Journal of Clinical Investigation, 2012, 122, 3731-3736.	3.9	123
86	SIRT2 Ablation Has No Effect on Tubulin Acetylation in Brain, Cholesterol Biosynthesis or the Progression of Huntington's Disease Phenotypes In Vivo. PLoS ONE, 2012, 7, e34805.	1.1	116
87	Fragments of HdhQ150 Mutant Huntingtin Form a Soluble Oligomer Pool That Declines with Aggregate Deposition upon Aging. PLoS ONE, 2012, 7, e44457.	1.1	21
88	Oral Administration of the Pimelic Diphenylamide HDAC Inhibitor HDACi 4b Is Unsuitable for Chronic Inhibition of HDAC Activity in the CNS In Vivo. PLoS ONE, 2012, 7, e44498.	1.1	34
89	A Brain-Permeable Small Molecule Reduces Neuronal Cholesterol by Inhibiting Activity of Sirtuin 2 Deacetylase. ACS Chemical Biology, 2011, 6, 540-546.	1.6	117
90	Hdac6 Knock-Out Increases Tubulin Acetylation but Does Not Modify Disease Progression in the R6/2 Mouse Model of Huntington's Disease. PLoS ONE, 2011, 6, e20696.	1.1	91

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91	Gastrointestinal dysfunction contributes to weight loss in Huntington's disease mice. Neurobiology of Disease, 2011, 44, 1-8.	2.1	88
92	The new science of ageing. Philosophical Transactions of the Royal Society B: Biological Sciences, 2011, 366, 6-8.	1.8	24
93	Altered chromatin architecture underlies progressive impairment of the heat shock response in mouse models of Huntington disease. Journal of Clinical Investigation, 2011, 121, 3306-3319.	3.9	151
94	The importance of integrating basic and clinical research toward the development of new therapies for Huntington disease. Journal of Clinical Investigation, 2011, 121, 476-483.	3.9	95
95	SAHA Decreases HDAC 2 and 4 Levels In Vivo and Improves Molecular Phenotypes in the R6/2 Mouse Model of Huntington's Disease. PLoS ONE, 2011, 6, e27746.	1.1	137
96	CalDAG-GEFI down-regulation in the striatum as a neuroprotective change in Huntington's disease. Human Molecular Genetics, 2010, 19, 1756-1765.	1.4	30
97	Identical oligomeric and fibrillar structures captured from the brains of R6/2 and knock-in mouse models of Huntington's disease. Human Molecular Genetics, 2010, 19, 65-78.	1.4	185
98	Proteolysis of Mutant Huntingtin Produces an Exon 1 Fragment That Accumulates as an Aggregated Protein in Neuronal Nuclei in Huntington Disease. Journal of Biological Chemistry, 2010, 285, 8808-8823.	1.6	259
99	SIRT2 inhibition achieves neuroprotection by decreasing sterol biosynthesis. Proceedings of the National Academy of Sciences of the United States of America, 2010, 107, 7927-7932.	3.3	304
100	Formation of Polyglutamine Inclusions in a Wide Range of Non-CNS Tissues in the HdhQ150 Knock-In Mouse Model of Huntington's Disease. PLoS ONE, 2009, 4, e8025.	1.1	131
101	Exendin-4 Improves Glycemic Control, Ameliorates Brain and Pancreatic Pathologies, and Extends Survival in a Mouse Model of Huntington's Disease. Diabetes, 2009, 58, 318-328.	0.3	160
102	A Large Number of Protein Expression Changes Occur Early in Life and Precede Phenotype Onset in a Mouse Model for Huntington Disease. Molecular and Cellular Proteomics, 2009, 8, 720-734.	2.5	66
103	IKK phosphorylates Huntingtin and targets it for degradation by the proteasome and lysosome. Journal of Cell Biology, 2009, 187, 1083-1099.	2.3	343
104	Systematic behavioral evaluation of Huntington's disease transgenic and knock-in mouse models. Neurobiology of Disease, 2009, 35, 319-336.	2.1	281
105	The polyubiquitin Ubc gene modulates histone H2A monoubiquitylation in the R6/2 mouse model of Huntington's disease. Journal of Cellular and Molecular Medicine, 2009, 13, 2645-2657.	1.6	23
106	The Ubiquitin-Proteasome Reporter GFPu Does Not Accumulate in Neurons of the R6/2 Transgenic Mouse Model of Huntington's Disease. PLoS ONE, 2009, 4, e5128.	1.1	43
107	Genetic Knock-Down of HDAC7 Does Not Ameliorate Disease Pathogenesis in the R6/2 Mouse Model of Huntington's Disease. PLoS ONE, 2009, 4, e5747.	1.1	61
108	Sensitive biochemical aggregate detection reveals aggregation onset before symptom development in cellular and murine models of Huntingtonâ€™s disease. Journal of Neurochemistry, 2008, 104, 846-858.	2.1	103

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109	Optimisation of region-specific reference gene selection and relative gene expression analysis methods for pre-clinical trials of Huntington's disease. Molecular Neurodegeneration, 2008, 3, 17.	4.4	48
110	Increased metabolism in the R6/2 mouse model of Huntingtonâ€™s disease. Neurobiology of Disease, 2008, 29, 41-51.	2.1	114
111	DNA instability in postmitotic neurons. Proceedings of the National Academy of Sciences of the United States of America, 2008, 105, 3467-3472.	3.3	184
112	A novel pathogenic pathway of immune activation detectable before clinical onset in Huntington's disease. Journal of Experimental Medicine, 2008, 205, 1869-1877.	4.2	559
113	Hsp27 overexpression in the R6/2 mouse model of Huntington's disease: chronic neurodegeneration does not induce Hsp27 activation. Human Molecular Genetics, 2007, 16, 1078-1090.	1.4	83
114	Mutant huntingtin's effects on striatal gene expression in mice recapitulate changes observed in human Huntington's disease brain and do not differ with mutant huntingtin length or wild-type huntingtin dosage. Human Molecular Genetics, 2007, 16, 1845-1861.	1.4	304
115	The HdhQ150/Q150 knock-in mouse model of HD and the R6/2 exon 1 model develop comparable and widespread molecular phenotypes. Brain Research Bulletin, 2007, 72, 83-97.	1.4	157
116	Proteomic Profiling of Plasma in Huntington's Disease Reveals Neuroinflammatory Activation and Biomarker Candidates. Journal of Proteome Research, 2007, 6, 2833-2840.	1.8	212
117	Global changes to the ubiquitin system in Huntington's disease. Nature, 2007, 448, 704-708.	13.7	478
118	Molecular Pathogenesis and Therapeutic Targets in Huntington's Disease. , 2006, , 223-249.		1
119	Metabolic Characterization of the R6/2 Transgenic Mouse Model of Huntington's Disease by High-Resolution MAS1H NMR Spectroscopy. Journal of Proteome Research, 2006, 5, 483-492.	1.8	119
120	Histone deacetylase inhibitors as therapeutics for polyglutamine disorders. Nature Reviews Neuroscience, 2006, 7, 784-796.	4.9	194
121	Mouse Models of Triplet Repeat Diseases. Molecular Biotechnology, 2006, 32, 147-158.	1.3	20
122	Evaluation of the benzothiazole aggregation inhibitors riluzole and PGL-135 as therapeutics for Huntington's disease. Neurobiology of Disease, 2006, 21, 228-236.	2.1	71
123	Early and transient alteration of adenosine A2A receptor signaling in a mouse model of Huntington disease. Neurobiology of Disease, 2006, 23, 44-53.	2.1	75
124	Elevated brain 3-hydroxykynurenine and quinolinate levels in Huntington disease mice. Neurobiology of Disease, 2006, 23, 190-197.	2.1	137
125	Proteasome impairment does not contribute to pathogenesis in R6/2 Huntington's disease mice: exclusion of proteasome activator REGÎ³ as a therapeutic target. Human Molecular Genetics, 2006, 15, 33-44.	1.4	91
126	BIOMEDICINE: One Misfolded Protein Allows Others to Sneak By. Science, 2006, 311, 1385-1386.	6.0	12

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127	Proteasome impairment does not contribute to pathogenesis in R6/2 Huntington's disease mice: exclusion of proteasome activator REGÎ³ as a therapeutic target. Human Molecular Genetics, 2006, 15, 665-665.	1.4	1
128	Progressive alterations in the hypothalamic-pituitary-adrenal axis in the R6/2 transgenic mouse model of Huntington's disease. Human Molecular Genetics, 2006, 15, 1713-1721.	1.4	122
129	Biomarkers for neurodegenerative diseases. Current Opinion in Neurology, 2005, 18, 698-705.	1.8	74
130	Reduction of GnRH and infertility in the R6/2 mouse model of Huntington's disease. European Journal of Neuroscience, 2005, 22, 1541-1546.	1.2	61
131	Polyglutamine expansion of huntingtin impairs its nuclear export. Nature Genetics, 2005, 37, 198-204.	9.4	153
132	The molecular genetics of Huntington disease â€" a history. Nature Reviews Genetics, 2005, 6, 766-773.	7.7	158
133	Contribution of nuclear and extranuclear polyQ to neurological phenotypes in mouse models of Huntington's disease. Human Molecular Genetics, 2005, 14, 3065-3078.	1.4	108
134	Dysfunction of the Cholesterol Biosynthetic Pathway in Huntington's Disease. Journal of Neuroscience, 2005, 25, 9932-9939.	1.7	236
135	A potent small molecule inhibits polyglutamine aggregation in Huntington's disease neurons and suppresses neurodegeneration in vivo. Proceedings of the National Academy of Sciences of the United States of America, 2005, 102, 892-897.	3.3	257
136	A human single-chain Fv intrabody preferentially targets amino-terminal huntingtin fragments in striatal models of Huntington's disease. Neurobiology of Disease, 2005, 19, 47-56.	2.1	48
137	Depletion of rabphilin 3A in a transgenic mouse model (R6/1) of Huntington's disease, a possible culprit in synaptic dysfunction. Neurobiology of Disease, 2005, 20, 673-684.	2.1	33
138	Mouse Models of Triplet Repeat Diseases. , 2004, 277, 003-016.		1
139	Monitoring Aggregate Formation in Organotypic Slice Cultures From Transgenic Mice. , 2004, 277, 161-172.		3
140	Huntingtin and the molecular pathogenesis of Huntington's disease. EMBO Reports, 2004, 5, 958-963.	2.0	429
141	Progressive decrease in chaperone protein levels in a mouse model of Huntington's disease and induction of stress proteins as a therapeutic approach. Human Molecular Genetics, 2004, 13, 1389-1405.	1.4	302
142	Minocycline and doxycycline are not beneficial in a model of Huntington's disease. Annals of Neurology, 2003, 54, 186-196.	2.8	153
143	Changes in GAD67 mRNA expression evidenced by in situ hybridization in the brain of R6/2 transgenic mice. Journal of Neurochemistry, 2003, 86, 1369-1378.	2.1	27
144	Inclusion formation in Huntington's disease R6/2 mouse muscle cultures. Journal of Neurochemistry, 2003, 87, 1-6.	2.1	41

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145	Complex alteration of NMDA receptors in transgenic Huntington's disease mouse brain: analysis of mRNA and protein expression, plasma membrane association, interacting proteins, and phosphorylation. Neurobiology of Disease, 2003, 14, 624-636.	2.1	92
146	Huntingtin aggregation and toxicity in Huntington's disease. Lancet, The, 2003, 361, 1642-1644.	6.3	470
147	Standardization and statistical approaches to therapeutic trials in the R6/2 mouse. Brain Research Bulletin, 2003, 61, 469-479.	1.4	129
148	Suberoylanilide hydroxamic acid, a histone deacetylase inhibitor, ameliorates motor deficits in a mouse model of Huntington's disease. Proceedings of the National Academy of Sciences of the United States of America, 2003, 100, 2041-2046.	3.3	805
149	Title is missing!. Current Opinion in Neurology, 2003, 16, 465-470.	1.8	6
150	Experimental therapeutics in Huntington's disease. Current Opinion in Neurology, 2003, 16, 465-470.	1.8	45
151	Alterations in the Mouse and Human Proteome Caused by Huntington's Disease. Molecular and Cellular Proteomics, 2002, 1, 366-375.	2.5	77
152	Abnormal Phosphorylation of Synapsin I Predicts a Neuronal Transmission Impairment in the R6/2 Huntington's Disease Transgenic Mice. Molecular and Cellular Neurosciences, 2002, 20, 638-648.	1.0	74
153	Environmental enrichment slows disease progression in R6/2 Huntington's disease mice. Annals of Neurology, 2002, 51, 235-242.	2.8	303
154	Arfaptin 2 regulates the aggregation of mutant huntingtin protein. Nature Cell Biology, 2002, 4, 240-245.	4.6	45
155	Impaired Glutamate Uptake in the R6 Huntington's Disease Transgenic Mice. Neurobiology of Disease, 2001, 8, 807-821.	2.1	271
156	Inhibition of Polyglutamine Aggregation in R6/2 HD Brain Slicesâ€"Complex Doseâ€"Response Profiles. Neurobiology of Disease, 2001, 8, 1017-1026.	2.1	69
157	Partial resistance to malonate-induced striatal cell death in transgenic mouse models of Huntington's disease is dependent on age and CAG repeat length. Journal of Neurochemistry, 2001, 78, 694-703.	2.1	53
158	Brain Neurotransmitter Deficits in Mice Transgenic for the Huntingtonâ€™s Disease Mutation. Journal of Neurochemistry, 2001, 72, 1773-1776.	2.1	84
159	Loss of cortical and thalamic neuronal tenascin-C expression in a transgenic mouse expressing exon 1 of the human Huntington disease gene. Journal of Comparative Neurology, 2001, 430, 485-500.	0.9	26
160	Exploiting expression. Nature, 2001, 413, 691-693.	13.7	17
161	The huntingtin interacting protein HIP1 is a clathrin and alpha-adaptin-binding protein involved in receptor-mediated endocytosis. Human Molecular Genetics, 2001, 10, 1807-1817.	1.4	139
162	Centrosome disorganization in fibroblast cultures derived from R6/2 Huntington's disease (HD) transgenic mice and HD patients. Human Molecular Genetics, 2001, 10, 2425-2435.	1.4	93

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163	Hunting in the calm before the storm. Nature Genetics, 2000, 25, 365-366.	9.4	6
164	In reverse gear. Nature, 2000, 404, 944-945.	13.7	6
165	Abnormal Synaptic Plasticity and Impaired Spatial Cognition in Mice Transgenic for Exon 1 of the Human Huntington's Disease Mutation. Journal of Neuroscience, 2000, 20, 5115-5123.	1.7	366
166	Nonapoptotic neurodegeneration in a transgenic mouse model of Huntington's disease. Proceedings of the National Academy of Sciences of the United States of America, 2000, 97, 8093-8097.	3.3	407
167	The Huntington's disease protein interacts with p53 and CREB-binding protein and represses transcription. Proceedings of the National Academy of Sciences of the United States of America, 2000, 97, 6763-6768.	3.3	966
168	Amyloid-like inclusions in Huntingtonâ€™s disease. Neuroscience, 2000, 100, 677-680.	1.1	93
169	Mitochondrial dysfunction and free radical damage in the Huntington R6/2 transgenic mouse. Annals of Neurology, 2000, 47, 80-6.	2.8	131
170	Transgenic Mouse Models of Huntingtonâ€™s Disease. , 2000, , 355-367.		1
171	Characterization of Progressive Motor Deficits in Mice Transgenic for the Human Huntingtonâ€™s Disease Mutation. Journal of Neuroscience, 1999, 19, 3248-3257.	1.7	864
172	Selective Discrimination Learning Impairments in Mice Expressing the Human Huntington's Disease Mutation. Journal of Neuroscience, 1999, 19, 10428-10437.	1.7	355
173	Formation of Polyglutamine Inclusions in Non-CNS Tissue. Human Molecular Genetics, 1999, 8, 813-822.	1.4	267
174	Ultrastructural localization and progressive formation of neuropil aggregates in Huntington's disease transgenic mice. Human Molecular Genetics, 1999, 8, 1227-1236.	1.4	182
175	Self-assembly of polyglutamine-containing huntingtin fragments into amyloid-like fibrils: Implications for Huntington's disease pathology. Proceedings of the National Academy of Sciences of the United States of America, 1999, 96, 4604-4609.	3.3	666
176	Transgenic models of Huntington'sdisease. Philosophical Transactions of the Royal Society B: Biological Sciences, 1999, 354, 963-969.	1.8	64
177	From neuronal inclusions to neurodegeneration: neuropathological investigation of a transgenic mouse model of Huntington'sdisease. Philosophical Transactions of the Royal Society B: Biological Sciences, 1999, 354, 971-979.	1.8	61
178	Altered neurotransmitter receptor expression in transgenic mouse models of Huntington's disease. Philosophical Transactions of the Royal Society B: Biological Sciences, 1999, 354, 981-989.	1.8	211
179	[43] Detection of polyglutamine aggregation in mouse models. Methods in Enzymology, 1999, 309, 687-701.	0.4	28
180	Molecular Pathology of Huntington's Disease: Animal Models and Nuclear Mechanisms. Neuroscientist, 1999, 5, 383-391.	2.6	3

#	Article	IF	Citations
181	Transgenic Mice in the Study of Polyglutamine Repeat Expansion Diseases. Brain Pathology, 1998, 8, 699-714.	2.1	63
182	A novel gene encoding an integral membrane protein is mutated in nephropathic cystinosis. Nature Genetics, 1998, 18, 319-324.	9.4	562
183	Striking changes in anxiety in Huntington's disease transgenic mice. Brain Research, 1998, 805, 234-240.	1.1	69
184	Are neuronal intranuclear inclusions the common neuropathology of triplet-repeat disorders with polyglutamine-repeat expansions?. Lancet, The, 1998, 351, 131-133.	6.3	173
185	SH3GL3 Associates with the Huntingtin Exon 1 Protein and Promotes the Formation of Polygln-Containing Protein Aggregates. Molecular Cell, 1998, 2, 427-436.	4.5	208
186	HAP1-huntingtin interactions do not contribute to the molecular pathology in Huntington's disease transgenic mice. FEBS Letters, 1998, 426, 229-232.	1.3	37
187	Striatal Transplantation in a Transgenic Mouse Model of Huntington's Disease. Experimental Neurology, 1998, 154, 31-40.	2.0	113
188	Intranuclear Neuronal Inclusions in Huntington's Disease and Dentatorubral and Pallidoluysian Atrophy: Correlation between the Density of Inclusions andIT15CAG Triplet Repeat Length. Neurobiology of Disease, 1998, 4, 387-397.	2.1	408
189	Altered brain neurotransmitter receptors in transgenic mice expressing a portion of an abnormal human Huntington disease gene. Proceedings of the National Academy of Sciences of the United States of America, 1998, 95, 6480-6485.	3.3	481
190	Polyglutamine expansion and Huntington's disease. Biochemical Society Transactions, 1998, 26, 471-475.	1.6	15
191	Aberrant Processing of the Fugu HD (FrHD) mRNA in Mouse Cells and in Transgenic Mice. Human Molecular Genetics, 1997, 6, 2141-2149.	1.4	21
192	Sequence comparison of human and yeast telomeres identifies structurally distinct subtelomeric domains. Human Molecular Genetics, 1997, 6, 1305-1313.	1.4	121
193	Transgenic models of Huntington's disease. Human Molecular Genetics, 1997, 6, 1633-1637.	1.4	97
194	Formation of Neuronal Intranuclear Inclusions Underlies the Neurological Dysfunction in Mice Transgenic for the HD Mutation. Cell, 1997, 90, 537-548.	13.5	2,105
195	Huntingtin-Encoded Polyglutamine Expansions Form Amyloid-like Protein Aggregates In Vitro and In Vivo. Cell, 1997, 90, 549-558.	13.5	1,224
196	Instability of highly expanded CAG repeats in mice transgenic for the Huntington's disease mutation. Nature Genetics, 1997, 15, 197-200.	9.4	302
197	Transgenic mouse models of neurodegenerative disease caused by CAG/polyglutamine expansions. Trends in Molecular Medicine, 1997, 3, 508-515.	2.6	28
198	Aggregation of Huntingtin in Neuronal Intranuclear Inclusions and Dystrophic Neurites in Brain. Science, 1997, 277, 1990-1993.	6.0	2,550

#	Article	IF	Citations
199	Exon trapping and sequence-based methods of gene finding in transcript mapping of human 4p 16.3. Somatic Cell and Molecular Genetics, 1997, 23, 413-427.	0.7	4
200	Identification of an HD patient with a (CAG) 180 repeat expansion and the propagation of highly expanded CAG repeats in lambda phage. Human Genetics, 1997, 99, 692-695.	1.8	56
201	Exon 1 of the HD Gene with an Expanded CAG Repeat Is Sufficient to Cause a Progressive Neurological Phenotype in Transgenic Mice. Cell, 1996, 87, 493-506.	13.5	2,892
202	Expanded glutamines and neurodegeneration - a gain of insight. BioEssays, 1996, 18, 175-178.	1.2	23
203	Transcript Map of the Human Chromosome 4p16.3 Consisting of 627 cDNA Clones Derived from 1 Mb of the Huntington's Disease Locus. DNA Research, 1996, 3, 239-255.	1.5	6
204	Distribution of trinucleotide repeat sequences across a 2 Mbp region containing the Huntington's disease gene. Human Molecular Genetics, 1994, 3, 73-78.	1.4	7
205	Trinucleotide repeat expansions and human genetic disease. BioEssays, 1994, 16, 277-284.	1.2	125
206	Structure and expression of the Huntington's disease gene: Evidence against simple inactivation due to an expanded CAG repeat. Somatic Cell and Molecular Genetics, 1994, 20, 27-38.	0.7	246
207	False-negative result for Huntington's disease mutation. Lancet, The, 1994, 343, 1232.	6.3	6
208	A cosmid contig and high resolution restriction map of the 2 megabase region containing the Huntington's disease gene. Nature Genetics, 1993, 4, 181-186.	9.4	102
209	Generation of high-density DNA markers from yeast artificial chromosome DNA by single unique primer-polymerase chain reaction. Genetic Analysis, Techniques and Applications, 1993, 10, 105-108.	1.5	2
210	Regional localization of the gene coding for human brain nitric oxide synthase (NOS1) to 12q24.2→24.31 by fluorescent in situ hybridization. Cytogenetic and Genome Research, 1993, 64, 62-63.	0.6	56
211	A novel gene containing a trinucleotide repeat that is expanded and unstable on Huntington's disease chromosomes. Cell, 1993, 72, 971-983.	13.5	7,960
212	A zinc-finger gene ZNF141 mapping at 4p16.3/D4S90 is a candidate gene for the Wolf-Hirschhorn (4p-) syndrome. Human Molecular Genetics, 1993, 2, 1571-1575.	1.4	48
213	The isolation of cDNAs within the Huntington disease region by hybridisation of yeast artificial chromosomes to a cDNA library. Human Molecular Genetics, 1993, 2, 305-309.	1.4	16
214	The Huntington disease geneâ€"still a needle in a haystack?. Human Molecular Genetics, 1993, 2, 343-347.	1.4	1
215	A novel G protein-coupled receptor kinase gene cloned from 4p16.3. Human Molecular Genetics, 1992, 1, 697-703.	1.4	100
216	The telomeric 60 kb of chromosome arm 4p is homologous to telomeric regions on 13p, 15p, 21p, and 22p. Genomics, 1992, 14, 350-356.	1.3	35

#	Article	IF	Citations
217	Radiation hybrid map spanning the huntington disease gene region of chromosome 4. Genomics, 1992, 13, 1040-1046.	1.3	20
218	Sequence-tagged sites (STSs) spanning 4p16.3 and the Huntington disease candidate region. Genomics, 1992, 13, 75-80.	1.3	16
219	The Huntington's disease candidate region exhibits many different haplotypes. Nature Genetics, 1992, 1, 99-103.	9.4	157
220	Characterization of a yeast artificial chromosome contig spanning the Huntington's disease gene candidate region. Nature Genetics, 1992, 1, 180-187.	9.4	71
221	Cloning of the Î±â€“adducin gene from the Huntington's disease candidate region of chromosome 4 by exon amplification. Nature Genetics, 1992, 2, 223-227.	9.4	47
222	New DNA markers in the Huntington's disease gene candidate region. Somatic Cell and Molecular Genetics, 1991, 17, 481-488.	0.7	27
223	Mapping of cosmid clones in Huntington's disease region of chromosome 4. Somatic Cell and Molecular Genetics, 1991, 17, 83-91.	0.7	46
224	The direct screening of cosmid libraries with YAC clones. Nucleic Acids Research, 1991, 19, 6651-6651.	6.5	34
225	The human homeobox gene HOX7 maps to chromosome 4p16.1 and may be implicated in Wolf-Hirschhorn syndrome. Human Genetics, 1990, 84, 473-6.	1.8	70
226	Physical maps of 4p16.3, the area expected to contain the Huntington disease mutation. Genomics, 1990, 6, 1-15.	1.3	87
227	A long-range restriction map encompassing the cystic fibrosis locus and its closely linked genetic markers. Genomics, 1988, 2, 337-345.	1.3	43
228	RFLP for pHM20 (D2S12), an anonymous DNA sequence localised to 2p23-2pter. Nucleic Acids Research, 1987, 15, 864-864.	6.5	0
229	Biochemical and genetic exclusion of calmodulin as the site of the basic defect in cystic fibrosis. Human Genetics, 1987, 76, 278-82.	1.8	23
230	A candidate for the cystic fibrosis locus isolated by selection for methylation-free islands. Nature, 1987, 326, 840-845.	13.7	364
231	Cystic fibrosis linkage exclusion data. Cytogenetic and Genome Research, 1986, 41, 62-63.	0.6	2
232	Isolation of a further anonymous informative DNA sequence from chromosome seven closely linked to cystic flbrosis. Nucleic Acids Research, 1986, 14, 1951-1956.	6.5	88
233	RELF for D4S12, an anonymous single copy genomic clone at 4pter-4q29 [HGM8 provisional no. D4S12]. Nucleic Acids Research, 1985, 13, 3016-3016.	6.5	2