Gillian P Bates

List of Publications by Year
in descending order

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		4383	2330
233	41,305	86	199
papers	citations	h-index	g-index

242	242	242	21250
all docs	docs citations	times ranked	citing authors

#	Article	IF	Citations
1	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
2	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
3	Aggregation of Huntingtin in Neuronal Intranuclear Inclusions and Dystrophic Neurites in Brain. Science, 1997, 277, 1990-1993.	6.0	2,550
4	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
5	Huntingtin-Encoded Polyglutamine Expansions Form Amyloid-like Protein Aggregates In Vitro and In Vivo. Cell, 1997, 90, 549-558.	13.5	1,224
6	Huntington disease. Nature Reviews Disease Primers, 2015, 1, 15005.	18.1	1,031
7	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
8	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
9	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
10	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
11	A novel gene encoding an integral membrane protein is mutated in nephropathic cystinosis. Nature Genetics, 1998, 18, 319-324.	9.4	562
12	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
13	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
14	Global changes to the ubiquitin system in Huntington's disease. Nature, 2007, 448, 704-708.	13.7	478
15	Huntingtin aggregation and toxicity in Huntington's disease. Lancet, The, 2003, 361, 1642-1644.	6.3	470
16	Huntingtin and the molecular pathogenesis of Huntington's disease. EMBO Reports, 2004, 5, 958-963.	2.0	429
17	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
18	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

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19	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
20	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
21	A candidate for the cystic fibrosis locus isolated by selection for methylation-free islands. Nature, 1987, 326, 840-845.	13.7	364
22	Selective Discrimination Learning Impairments in Mice Expressing the Human Huntington's Disease Mutation. Journal of Neuroscience, 1999, 19, 10428-10437.	1.7	355
23	IKK phosphorylates Huntingtin and targets it for degradation by the proteasome and lysosome. Journal of Cell Biology, 2009, 187, 1083-1099.	2.3	343
24	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
25	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
26	Environmental enrichment slows disease progression in R6/2 Huntington's disease mice. Annals of Neurology, 2002, 51, 235-242.	2.8	303
27	Instability of highly expanded CAG repeats in mice transgenic for the Huntington's disease mutation. Nature Genetics, 1997, 15, 197-200.	9.4	302
28	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
29	Systematic behavioral evaluation of Huntington's disease transgenic and knock-in mouse models. Neurobiology of Disease, 2009, 35, 319-336.	2.1	281
30	Impaired Glutamate Uptake in the R6 Huntington's Disease Transgenic Mice. Neurobiology of Disease, 2001, 8, 807-821.	2.1	271
31	Formation of Polyglutamine Inclusions in Non-CNS Tissue. Human Molecular Genetics, 1999, 8, 813-822.	1.4	267
32	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
33	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
34	UBQLN2 Mediates Autophagy-Independent Protein Aggregate Clearance by the Proteasome. Cell, 2016, 166, 935-949.	13.5	248
35	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
36	Dysfunction of the Cholesterol Biosynthetic Pathway in Huntington's Disease. Journal of Neuroscience, 2005, 25, 9932-9939.	1.7	236

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37	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
38	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
39	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
40	Histone deacetylase inhibitors as therapeutics for polyglutamine disorders. Nature Reviews Neuroscience, 2006, 7, 784-796.	4.9	194
41	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
42	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
43	Ultrastructural localization and progressive formation of neuropil aggregates in Huntington's disease transgenic mice. Human Molecular Genetics, 1999, 8, 1227-1236.	1.4	182
44	Are neuronal intranuclear inclusions the common neuropathology of triplet-repeat disorders with polyglutamine-repeat expansions?. Lancet, The, 1998, 351, 131-133.	6.3	173
45	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
46	The molecular genetics of Huntington disease â€" a history. Nature Reviews Genetics, 2005, 6, 766-773.	7.7	158
47	The Huntington's disease candidate region exhibits many different haplotypes. Nature Genetics, 1992, 1, 99-103.	9.4	157
48	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
49	Minocycline and doxycycline are not beneficial in a model of Huntington's disease. Annals of Neurology, 2003, 54, 186-196.	2.8	153
50	Polyglutamine expansion of huntingtin impairs its nuclear export. Nature Genetics, 2005, 37, 198-204.	9.4	153
51	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
52	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
53	The pathogenic exon 1 HTT protein is produced by incomplete splicing in Huntingtonâ€™s disease patients. Scientific Reports, 2017, 7, 1307.	1.6	150
54	HTT-lowering reverses Huntingtonâ€™s disease immune dysfunction caused by NFÎºB pathway dysregulation. Brain, 2014, 137, 819-833.	3.7	147

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55	HDAC4 Reduction: A Novel Therapeutic Strategy to Target Cytoplasmic Huntingtin and Ameliorate Neurodegeneration. PLoS Biology, 2013, 11, e1001717.	2.6	143
56	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
57	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
58	Elevated brain 3-hydroxykynurenine and quinolinate levels in Huntington disease mice. Neurobiology of Disease, 2006, 23, 190-197.	2.1	137
59	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
60	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
61	Mitochondrial dysfunction and free radical damage in the Huntington R6/2 transgenic mouse. Annals of Neurology, 2000, 47, 80-6.	2.8	131
62	Standardization and statistical approaches to therapeutic trials in the R6/2 mouse. Brain Research Bulletin, 2003, 61, 469-479.	1.4	129
63	Treating the whole body in Huntington's disease. Lancet Neurology, The, 2015, 14, 1135-1142.	4.9	126
64	Trinucleotide repeat expansions and human genetic disease. BioEssays, 1994, 16, 277-284.	1.2	125
65	Mutant huntingtin fragmentation in immune cells tracks Huntingtonâ€™s disease progression. Journal of Clinical Investigation, 2012, 122, 3731-3736.	3.9	123
66	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
67	Sequence comparison of human and yeast telomeres identifies structurally distinct subtelomeric domains. Human Molecular Genetics, 1997, 6, 1305-1313.	1.4	121
68	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
69	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
70	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
71	Increased metabolism in the R6/2 mouse model of Huntingtonâ€™s disease. Neurobiology of Disease, 2008, 29, 41-51.	2.1	114
72	Striatal Transplantation in a Transgenic Mouse Model of Huntington's Disease. Experimental Neurology, 1998, 154, 31-40.	2.0	113

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73	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
74	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
75	Suppression of protein aggregation by chaperone modification of high molecular weight complexes. Brain, 2012, 135, 1180-1196.	3.7	103
76	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
77	A novel G protein-coupled receptor kinase gene cloned from 4p16.3. Human Molecular Genetics, 1992, 1, 697-703.	1.4	100
78	Transgenic models of Huntington's disease. Human Molecular Genetics, 1997, 6, 1633-1637.	1.4	97
79	SUMO-2 and PIAS1 Modulate Insoluble Mutant Huntingtin Protein Accumulation. Cell Reports, 2013, 4, 362-375.	2.9	97
80	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
81	Amyloid-like inclusions in Huntingtonâ€™s disease. Neuroscience, 2000, 100, 677-680.	1.1	93
82	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
83	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
84	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
85	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
86	Isolation of a further anonymous informative DNA sequence from chromosome seven closely linked to cystic fibrosis. Nucleic Acids Research, 1986, 14, 1951-1956.	6.5	88
87	Gastrointestinal dysfunction contributes to weight loss in Huntington's disease mice. Neurobiology of Disease, 2011, 44, 1-8.	2.1	88
88	Physical maps of 4p16.3, the area expected to contain the Huntington disease mutation. Genomics, 1990, 6, 1-15.	1.3	87
89	Brain Neurotransmitter Deficits in Mice Transgenic for the Huntingtonâ€™s Disease Mutation. Journal of Neurochemistry, 2001, 72, 1773-1776.	2.1	84
90	Frequency of nuclear mutant huntingtin inclusion formation in neurons and glia is cellâ€typeâ€specific. Glia, 2017, 65, 50-61.	2.5	84

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91	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
92	Dysfunction of the CNS-Heart Axis in Mouse Models of Huntington's Disease. PLoS Genetics, 2014, 10, e1004550.	1.5	83
93	Alterations in the Mouse and Human Proteome Caused by Huntington's Disease. Molecular and Cellular Proteomics, 2002, 1, 366-375.	2.5	77
94	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
95	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
96	Biomarkers for neurodegenerative diseases. Current Opinion in Neurology, 2005, 18, 698-705.	1.8	74
97	Characterization of a yeast artificial chromosome contig spanning the Huntington's disease gene candidate region. Nature Genetics, 1992, 1, 180-187.	9.4	71
98	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
99	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
100	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
101	Striking changes in anxiety in Huntington's disease transgenic mice. Brain Research, 1998, 805, 234-240.	1.1	69
102	Inhibition of Polyglutamine Aggregation in R6/2 HD Brain Slicesâ€"Complex Doseâ€"Response Profiles. Neurobiology of Disease, 2001, 8, 1017-1026.	2.1	69
103	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
104	Transgenic models of Huntington'sdisease. Philosophical Transactions of the Royal Society B: Biological Sciences, 1999, 354, 963-969.	1.8	64
105	Transgenic Mice in the Study of Polyglutamine Repeat Expansion Diseases. Brain Pathology, 1998, 8, 699-714.	2.1	63
106	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
107	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
108	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

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109	Mouse Models of Huntingtonâ€™s Disease. Methods in Molecular Biology, 2018, 1780, 97-120.	0.4	57
110	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
111	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
112	HDAC4-Myogenin Axis As an Important Marker of HD-Related Skeletal Muscle Atrophy. PLoS Genetics, 2015, 11, e1005021.	1.5	56
113	Regulatory mechanisms of incomplete huntingtin mRNA splicing. Nature Communications, 2018, 9, 3955.	5.8	55
114	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
115	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
116	Aberrantly splicedHTT,a new player in Huntingtonâ€™s disease pathogenesis. RNA Biology, 2013, 10, 1647-1652.	1.5	50
117	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
118	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
119	A common gene expression signature in Huntingtonâ€™s disease patient brain regions. BMC Medical Genomics, 2014, 7, 60.	0.7	49
120	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
121	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
122	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
123	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
124	Stall in Canonical Autophagy-Lysosome Pathways Prompts Nucleophagy-Based Nuclear Breakdown in Neurodegeneration. Current Biology, 2017, 27, 3626-3642.e6.	1.8	47
125	Mapping of cosmid clones in Huntington's disease region of chromosome 4. Somatic Cell and Molecular Genetics, 1991, 17, 83-91.	0.7	46
126	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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127	Arfaptin 2 regulates the aggregation of mutant huntingtin protein. Nature Cell Biology, 2002, 4, 240-245.	4.6	45
128	Experimental therapeutics in Huntington's disease. Current Opinion in Neurology, 2003, 16, 465-470.	1.8	45
129	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
130	Dynamic recruitment of active proteasomes into polyglutamine initiated inclusion bodies. FEBS Letters, 2014, 588, 151-159.	1.3	44
131	A long-range restriction map encompassing the cystic fibrosis locus and its closely linked genetic markers. Genomics, 1988, 2, 337-345.	1.3	43
132	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
133	Inclusion formation in Huntington's disease R6/2 mouse muscle cultures. Journal of Neurochemistry, 2003, 87, 1-6.	2.1	41
134	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
135	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
136	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
137	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
138	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
139	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
140	The direct screening of cosmid libraries with YAC clones. Nucleic Acids Research, 1991, 19, 6651-6651.	6.5	34
141	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
142	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
143	TBK1 phosphorylates mutant Huntingtin and suppresses its aggregation and toxicity in Huntington's disease models. EMBO Journal, 2020, 39, e104671.	3.5	34
144	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

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145	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
146	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
147	Meso scale discovery-based assays for the detection of aggregated huntingtin. PLoS ONE, 2019, 14, e0213521.	1.1	31
148	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
149	HDAC4 Does Not Act as a Protein Deacetylase in the Postnatal Murine Brain In Vivo. PLoS ONE, 2013, 8, e80849.	1.1	30
150	Uninterrupted CAG repeat drives striatum-selective transcriptionopathy and nuclear pathogenesis in human Huntingtin BAC mice. Neuron, 2022, 110, 1173-1192.e7.	3.8	30
151	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
152	The Huntington's Disease-Related Cardiomyopathy Prevents a Hypertrophic Response in the R6/2 Mouse Model. PLoS ONE, 2014, 9, e108961.	1.1	29
153	Transgenic mouse models of neurodegenerative disease caused by CAG/polyglutamine expansions. Trends in Molecular Medicine, 1997, 3, 508-515.	2.6	28
154	[43] Detection of polyglutamine aggregation in mouse models. Methods in Enzymology, 1999, 309, 687-701.	0.4	28
155	New DNA markers in the Huntington's disease gene candidate region. Somatic Cell and Molecular Genetics, 1991, 17, 481-488.	0.7	27
156	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
157	Myostatin inhibition prevents skeletal muscle pathophysiology in Huntingtonâ€™s disease mice. Scientific Reports, 2017, 7, 14275.	1.6	27
158	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
159	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
160	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
161	The new science of ageing. Philosophical Transactions of the Royal Society B: Biological Sciences, 2011, 366, 6-8.	1.8	24
162	Systematic interaction network filtering identifies CRMP1 as a novel suppressor of huntingtin misfolding and neurotoxicity. Genome Research, 2015, 25, 701-713.	2.4	24

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163	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
164	Expanded glutamines and neurodegeneration - a gain of insight. BioEssays, 1996, 18, 175-178.	1.2	23
165	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
166	MEF2 impairment underlies skeletal muscle atrophy in polyglutamine disease. Acta Neuropathologica, 2020, 140, 63-80.	3.9	23
167	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
168	Disruption to schizophrenia-associated gene Fez1 in the hippocampus of HDAC11 knockout mice. Scientific Reports, 2017, 7, 11900.	1.6	21
169	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
170	Radiation hybrid map spanning the huntington disease gene region of chromosome 4. Genomics, 1992, 13, 1040-1046.	1.3	20
171	Mouse Models of Triplet Repeat Diseases. Molecular Biotechnology, 2006, 32, 147-158.	1.3	20
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