Gillian P Bates

List of Publications by Citations

219 papers	35,424 citations	82 h-index	188 g-index
242 ext. papers	38,317 ext. citations	10.6 avg, IF	7.15 L-index

#	Paper	IF	Citations
219	A novel gene containing a trinucleotide repeat that is expanded and unstable on Huntington's disease chromosomes. The Huntington's Disease Collaborative Research Group. *Cell*, **1993**, 72, 971-83	56.2	6854
218	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	56.2	2587
217	Aggregation of huntingtin in neuronal intranuclear inclusions and dystrophic neurites in brain. *Science*, **1997**, 277, 1990-3	33.3	2213
216	Formation of neuronal intranuclear inclusions underlies the neurological dysfunction in mice transgenic for the HD mutation. *Cell*, **1997**, 90, 537-48	56.2	1918
215	Huntingtin-encoded polyglutamine expansions form amyloid-like protein aggregates in vitro and in vivo. *Cell*, **1997**, 90, 549-58	56.2	1114
214	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, 6763-8	11.5	870
213	Characterization of progressive motor deficits in mice transgenic for the human Huntington's disease mutation. *Journal of Neuroscience*, **1999**, 19, 3248-57	6.6	784
212	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, 2041-6	11.5	734
211	Huntington disease. *Nature Reviews Disease Primers*, **2015**, 1, 15005	51.1	672
210	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, 4604-9	11.5	584
209	A novel gene encoding an integral membrane protein is mutated in nephropathic cystinosis. *Nature Genetics*, **1998**, 18, 319-24	36.3	458
208	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, 6480-5	11.5	450
207	Global changes to the ubiquitin system in Huntington's disease. *Nature*, **2007**, 448, 704-8	50.4	444
206	A novel pathogenic pathway of immune activation detectable before clinical onset in Huntington's disease. *Journal of Experimental Medicine*, **2008**, 205, 1869-77	16.6	437
205	Huntingtin aggregation and toxicity in Huntington's disease. *Lancet, The*, **2003**, 361, 1642-4	40	422
204	Intranuclear neuronal inclusions in Huntington's disease and dentatorubral and pallidoluysian atrophy: correlation between the density of inclusions and IT15 CAG triplet repeat length. *Neurobiology of Disease*, **1998**, 4, 387-97	7.5	373
203	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, 8093-7	11.5	369

202	Huntingtin and the molecular pathogenesis of Huntington's disease. Fourth in molecular medicine review series. *EMBO Reports*, **2004**, 5, 958-63	6.5	365
201	A candidate for the cystic fibrosis locus isolated by selection for methylation-free islands. *Nature*, **1987**, 326, 840-5	50.4	341
200	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, 5115-23	6.6	333
199	Selective discrimination learning impairments in mice expressing the human Huntington's disease mutation. *Journal of Neuroscience*, **1999**, 19, 10428-37	6.6	330
198	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, 2366-70	11.5	310
197	IKK phosphorylates Huntingtin and targets it for degradation by the proteasome and lysosome. *Journal of Cell Biology*, **2009**, 187, 1083-99	7.3	287
196	Environmental enrichment slows disease progression in R6/2 Huntington's disease mice. *Annals of Neurology*, **2002**, 51, 235-42	9.4	282
195	Instability of highly expanded CAG repeats in mice transgenic for the Huntington's disease mutation. *Nature Genetics*, **1997**, 15, 197-200	36.3	275
194	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, 1845-61	5.6	271
193	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, 1389-405	5.6	261
192	SIRT2 inhibition achieves neuroprotection by decreasing sterol biosynthesis. *Proceedings of the National Academy of Sciences of the United States of America*, **2010**, 107, 7927-32	11.5	255
191	Systematic behavioral evaluation of Huntington's disease transgenic and knock-in mouse models. *Neurobiology of Disease*, **2009**, 35, 319-36	7.5	244
190	Formation of polyglutamine inclusions in non-CNS tissue. *Human Molecular Genetics*, **1999**, 8, 813-22	5.6	243
189	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, 892-7	11.5	240
188	Impaired glutamate uptake in the R6 Huntington's disease transgenic mice. *Neurobiology of Disease* , **2001**, 8, 807-21	7.5	233
187	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, 8808-23	5.4	219
186	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		210
185	Dysfunction of the cholesterol biosynthetic pathway in Huntington's disease. *Journal of Neuroscience*, **2005**, 25, 9932-9	6.6	195

184	SH3GL3 associates with the Huntingtin exon 1 protein and promotes the formation of polygln-containing protein aggregates. *Molecular Cell*, **1998**, 2, 427-36	17.6	189
183	UBQLN2 Mediates Autophagy-Independent Protein Aggregate Clearance by the Proteasome. *Cell*, **2016**, 166, 935-949	56.2	186
182	Altered neurotransmitter receptor expression in transgenic mouse models of Huntington's disease. *Philosophical Transactions of the Royal Society B: Biological Sciences*, **1999**, 354, 981-9	5.8	185
181	Histone deacetylase inhibitors as therapeutics for polyglutamine disorders. *Nature Reviews Neuroscience*, **2006**, 7, 784-96	13.5	176
180	Proteomic profiling of plasma in Huntington's disease reveals neuroinflammatory activation and biomarker candidates. *Journal of Proteome Research*, **2007**, 6, 2833-40	5.6	173
179	Are neuronal intranuclear inclusions the common neuropathology of triplet-repeat disorders with polyglutamine-repeat expansions?. *Lancet, The*, **1998**, 351, 131-3	40	163
178	Ultrastructural localization and progressive formation of neuropil aggregates in Huntington's disease transgenic mice. *Human Molecular Genetics*, **1999**, 8, 1227-36	5.6	152
177	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	5.6	151
176	DNA instability in postmitotic neurons. *Proceedings of the National Academy of Sciences of the United States of America*, **2008**, 105, 3467-72	11.5	149
175	Minocycline and doxycycline are not beneficial in a model of Huntington's disease. *Annals of Neurology*, **2003**, 54, 186-96	9.4	145
174	The Huntington's disease candidate region exhibits many different haplotypes. *Nature Genetics*, **1992**, 1, 99-103	36.3	142
173	The Hdh(Q150/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	3.9	140
172	Exendin-4 improves glycemic control, ameliorates brain and pancreatic pathologies, and extends survival in a mouse model of Huntington's disease. *Diabetes*, **2009**, 58, 318-28	0.9	135
171	History of genetic disease: the molecular genetics of Huntington disease - a history. *Nature Reviews Genetics*, **2005**, 6, 766-73	30.1	132
170	Altered chromatin architecture underlies progressive impairment of the heat shock response in mouse models of Huntington disease. *Journal of Clinical Investigation*, **2011**, 121, 3306-19	15.9	130
169	Polyglutamine expansion of huntingtin impairs its nuclear export. *Nature Genetics*, **2005**, 37, 198-204	36.3	129
168	Elevated brain 3-hydroxykynurenine and quinolinate levels in Huntington disease mice. *Neurobiology of Disease*, **2006**, 23, 190-7	7.5	125
167	Standardization and statistical approaches to therapeutic trials in the R6/2 mouse. *Brain Research Bulletin*, **2003**, 61, 469-79	3.9	125

166	The huntingtin interacting protein HIP1 is a clathrin and alpha-adaptin-binding protein involved in receptor-mediated endocytosis. *Human Molecular Genetics*, **2001**, 10, 1807-17	5.6	118
165	HDAC4 reduction: a novel therapeutic strategy to target cytoplasmic huntingtin and ameliorate neurodegeneration. *PLoS Biology*, **2013**, 11, e1001717	9.7	117
164	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	3.7	117
163	Mitochondrial dysfunction and free radical damage in the Huntington R6/2 transgenic mouse. *Annals of Neurology*, **2000**, 47, 80-6	9.4	114
162	Targeting H3K4 trimethylation in Huntington disease. *Proceedings of the National Academy of Sciences of the United States of America*, **2013**, 110, E3027-36	11.5	112
161	Progressive alterations in the hypothalamic-pituitary-adrenal axis in the R6/2 transgenic mouse model of Huntington's disease. *Human Molecular Genetics*, **2006**, 15, 1713-21	5.6	110
160	HTT-lowering reverses Huntington's disease immune dysfunction caused by NFB pathway dysregulation. *Brain*, **2014**, 137, 819-33	11.2	109
159	Trinucleotide repeat expansions and human genetic disease. *BioEssays*, **1994**, 16, 277-84	4.1	107
158	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	3.7	106
157	Increased metabolism in the R6/2 mouse model of Huntington's disease. *Neurobiology of Disease*, **2008**, 29, 41-51	7.5	105
156	Striatal transplantation in a transgenic mouse model of Huntington's disease. *Experimental Neurology*, **1998**, 154, 31-40	5.7	105
155	A brain-permeable small molecule reduces neuronal cholesterol by inhibiting activity of sirtuin 2 deacetylase. *ACS Chemical Biology*, **2011**, 6, 540-6	4.9	99
154	Metabolic characterization of the R6/2 transgenic mouse model of Huntington's disease by high-resolution MAS 1H NMR spectroscopy. *Journal of Proteome Research*, **2006**, 5, 483-92	5.6	99
153	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	3.7	98
152	Mutant huntingtin fragmentation in immune cells tracks Huntington's disease progression. *Journal of Clinical Investigation*, **2012**, 122, 3731-6	15.9	97
151	Treating the whole body in Huntington's disease. *Lancet Neurology, The*, **2015**, 14, 1135-42	24.1	96
150	A novel G protein-coupled receptor kinase gene cloned from 4p16.3. *Human Molecular Genetics*, **1992**, 1, 697-703	5.6	95
149	A cosmid contig and high resolution restriction map of the 2 megabase region containing the Huntington's disease gene. *Nature Genetics*, **1993**, 4, 181-6	36.3	93

148	Contribution of nuclear and extranuclear polyQ to neurological phenotypes in mouse models of Huntington's disease. *Human Molecular Genetics*, **2005**, 14, 3065-78	5.6	92
147	Sequence comparison of human and yeast telomeres identifies structurally distinct subtelomeric domains. *Human Molecular Genetics*, **1997**, 6, 1305-13	5.6	91
146	The pathogenic exon 1 HTT protein is produced by incomplete splicing in Huntington's disease patients. *Scientific Reports*, **2017**, 7, 1307	4.9	89
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, 624-36	7.5	89
144	Sensitive biochemical aggregate detection reveals aggregation onset before symptom development in cellular and murine models of Huntington's disease. *Journal of Neurochemistry*, **2008**, 104, 846-58	6	88
143	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	17.6	87
142	Proteasome impairment does not contribute to pathogenesis in R6/2 Huntington's disease mice: exclusion of proteasome activator REGgamma as a therapeutic target. *Human Molecular Genetics*, **2006**, 15, 33-44	5.6	86
141	Centrosome disorganization in fibroblast cultures derived from R6/2 Huntington's disease (HD) transgenic mice and HD patients. *Human Molecular Genetics*, **2001**, 10, 2425-35	5.6	86
140	Isolation of a further anonymous informative DNA sequence from chromosome seven closely linked to cystic fibrosis. *Nucleic Acids Research*, **1986**, 14, 1951-6	20.1	86
139	Suppression of protein aggregation by chaperone modification of high molecular weight complexes. *Brain*, **2012**, 135, 1180-96	11.2	85
138	Amyloid-like inclusions in Huntington's disease. *Neuroscience*, **2000**, 100, 677-80	3.9	83
137	Physical maps of 4p16.3, the area expected to contain the Huntington disease mutation. *Genomics*, **1990**, 6, 1-15	4.3	79
136	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	3.7	77
135	The importance of integrating basic and clinical research toward the development of new therapies for Huntington disease. *Journal of Clinical Investigation*, **2011**, 121, 476-83	15.9	77
134	Hsp27 overexpression in the R6/2 mouse model of Huntington's disease: chronic neurodegeneration does not induce Hsp27 activation. *Human Molecular Genetics*, **2007**, 16, 1078-90	5.6	76
133	Transgenic models of Huntington's disease. *Human Molecular Genetics*, **1997**, 6, 1633-7	5.6	72
132	Brain neurotransmitter deficits in mice transgenic for the Huntington's disease mutation. *Journal of Neurochemistry*, **1999**, 72, 1773-6	6	72
131	SUMO-2 and PIAS1 modulate insoluble mutant huntingtin protein accumulation. *Cell Reports*, **2013**, 4, 362-75	10.6	68

130	Early and transient alteration of adenosine A2A receptor signaling in a mouse model of Huntington disease. *Neurobiology of Disease*, **2006**, 23, 44-53	7.5	68
129	Characterization of a yeast artificial chromosome contig spanning the Huntington's disease gene candidate region. *Nature Genetics*, **1992**, 1, 180-7	36.3	67
128	Alterations in the mouse and human proteome caused by Huntington's disease. *Molecular and Cellular Proteomics*, **2002**, 1, 366-75	7.6	66
127	Inhibition of polyglutamine aggregation in R6/2 HD brain slices-complex dose-response profiles. *Neurobiology of Disease*, **2001**, 8, 1017-26	7.5	66
126	Dysfunction of the CNS-heart axis in mouse models of Huntington's disease. *PLoS Genetics*, **2014**, 10, e1004550	6	65
125	Evaluation of the benzothiazole aggregation inhibitors riluzole and PGL-135 as therapeutics for Huntington's disease. *Neurobiology of Disease*, **2006**, 21, 228-36	7.5	63
124	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, 638-48	4.8	62
123	Striking changes in anxiety in Huntington's disease transgenic mice. *Brain Research*, **1998**, 805, 234-40	3.7	61
122	Gastrointestinal dysfunction contributes to weight loss in Huntington's disease mice. *Neurobiology of Disease*, **2011**, 44, 1-8	7.5	60
121	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, 720-34	7.6	59
120	TR-FRET-based duplex immunoassay reveals an inverse correlation of soluble and aggregated mutant huntingtin in huntington's disease. *Chemistry and Biology*, **2012**, 19, 264-75		58
119	Transgenic models of Huntington's disease. *Philosophical Transactions of the Royal Society B: Biological Sciences*, **1999**, 354, 963-9	5.8	58
118	The human homeobox gene HOX7 maps to chromosome 4p16.1 and may be implicated in Wolf-Hirschhorn syndrome. *Human Genetics*, **1990**, 84, 473-6	6.3	58
117	Frequency of nuclear mutant huntingtin inclusion formation in neurons and glia is cell-type-specific. *Glia*, **2017**, 65, 50-61	9	56
116	Biomarkers for neurodegenerative diseases. *Current Opinion in Neurology*, **2005**, 18, 698-705	7.1	55
115	Reduction of GnRH and infertility in the R6/2 mouse model of Huntington's disease. *European Journal of Neuroscience*, **2005**, 22, 1541-6	3.5	54
114	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, 62-3	1.9	53
113	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	3.7	53

112	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, 692-5	6.3	52
111	Transgenic mice in the study of polyglutamine repeat expansion diseases. *Brain Pathology*, **1998**, 8, 699-714	7	49
110	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	5.8	48
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	19	46
108	A human single-chain Fv intrabody preferentially targets amino-terminal Huntingtin's fragments in striatal models of Huntington's disease. *Neurobiology of Disease*, **2005**, 19, 47-56	7.5	45
107	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	6	45
106	HDAC4-myogenin axis as an important marker of HD-related skeletal muscle atrophy. *PLoS Genetics* , **2015**, 11, e1005021	6	44
105	Genetic knock-down of HDAC3 does not modify disease-related phenotypes in a mouse model of Huntington's disease. *PLoS ONE*, **2012**, 7, e31080	3.7	44
104	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, 1571-5	5.6	44
103	Arfaptin 2 regulates the aggregation of mutant huntingtin protein. *Nature Cell Biology*, **2002**, 4, 240-5	23.4	43
102	Cloning of the alpha-adducin gene from the Huntington's disease candidate region of chromosome 4 by exon amplification. *Nature Genetics*, **1992**, 2, 223-7	36.3	43
101	Mapping of cosmid clones in Huntington's disease region of chromosome 4. *Somatic Cell and Molecular Genetics*, **1991**, 17, 83-91		42
100	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	3.7	41
99	Dynamic recruitment of active proteasomes into polyglutamine initiated inclusion bodies. *FEBS Letters*, **2014**, 588, 151-9	3.8	40
98	Aberrantly spliced HTT, a new player in Huntington's disease pathogenesis. *RNA Biology*, **2013**, 10, 1647-53	4.8	40
97	A long-range restriction map encompassing the cystic fibrosis locus and its closely linked genetic markers. *Genomics*, **1988**, 2, 337-45	4.3	38
96	Characterisation of immune cell function in fragment and full-length Huntington's disease mouse models. *Neurobiology of Disease*, **2015**, 73, 388-98	7.5	37
95	Experimental therapeutics in Huntington's disease: are models useful for therapeutic trials?. *Current Opinion in Neurology*, **2003**, 16, 465-70	7.1	37

94	Inclusion formation in Huntington's disease R6/2 mouse muscle cultures. *Journal of Neurochemistry*, **2003**, 87, 1-6	6	35
93	Downregulation of cannabinoid receptor 1 from neuropeptide Y interneurons in the basal ganglia of patients with Huntington's disease and mouse models. *European Journal of Neuroscience*, **2013**, 37, 429-40	3.5	34
92	A common gene expression signature in Huntington's disease patient brain regions. *BMC Medical Genomics*, **2014**, 7, 60	3.7	34
91	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	3.7	34
90	Mouse Models of Huntington's Disease. *Methods in Molecular Biology*, **2018**, 1780, 97-120	1.4	34
89	Stall in Canonical Autophagy-Lysosome Pathways Prompts Nucleophagy-Based Nuclear Breakdown in Neurodegeneration. *Current Biology*, **2017**, 27, 3626-3642.e6	6.3	33
88	Implantation of undifferentiated and pre-differentiated human neural stem cells in the R6/2 transgenic mouse model of Huntington's disease. *BMC Neuroscience*, **2012**, 13, 97	3.2	33
87	mHTT Seeding Activity: A Marker of Disease Progression and Neurotoxicity in Models of Huntington's Disease. *Molecular Cell*, **2018**, 71, 675-688.e6	17.6	33
86	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	3.7	31
85	The direct screening of cosmid libraries with YAC clones. *Nucleic Acids Research*, **1991**, 19, 6651	20.1	30
84	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, 673-84	7.5	29
83	The telomeric 60 kb of chromosome arm 4p is homologous to telomeric regions on 13p, 15p, 21p, and 22p. *Genomics*, **1992**, 14, 350-6	4.3	29
82	CalDAG-GEFI down-regulation in the striatum as a neuroprotective change in Huntington's disease. *Human Molecular Genetics*, **2010**, 19, 1756-65	5.6	28
81	HAP1-huntingtin interactions do not contribute to the molecular pathology in Huntington's disease transgenic mice. *FEBS Letters*, **1998**, 426, 229-32	3.8	28
80	HDAC4 does not act as a protein deacetylase in the postnatal murine brain in vivo. *PLoS ONE*, **2013**, 8, e80849	3.7	27
79	Regulatory mechanisms of incomplete huntingtin mRNA splicing. *Nature Communications*, **2018**, 9, 3955	17.4	27
78	Contesting the dogma of an age-related heat shock response impairment: implications for cardiac-specific age-related disorders. *Human Molecular Genetics*, **2014**, 23, 3641-56	5.6	26
77	Detection of polyglutamine aggregation in mouse models. *Methods in Enzymology*, **1999**, 309, 687-701	1.7	26

76	New DNA markers in the Huntington's disease gene candidate region. *Somatic Cell and Molecular Genetics*, **1991**, 17, 481-8		25
75	Correlations of behavioral deficits with brain pathology assessed through longitudinal MRI and histopathology in the R6/1 mouse model of Huntington's disease. *PLoS ONE*, **2013**, 8, e84726	3.7	23
74	Changes in GAD67 mRNA expression evidenced by in situ hybridization in the brain of R6/2 transgenic mice. *Journal of Neurochemistry*, **2003**, 86, 1369-78	6	23
73	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	3.4	23
72	Expanded glutamines and neurodegeneration--a gain of insight. *BioEssays*, **1996**, 18, 175-8	4.1	23
71	Phenotype onset in Huntington's disease knock-in mice is correlated with the incomplete splicing of the mutant huntingtin gene. *Journal of Neuroscience Research*, **2019**, 97, 1590-1605	4.4	21
70	Transgenic mouse models of neurodegenerative disease caused by CAG/polyglutamine expansions. *Trends in Molecular Medicine*, **1997**, 3, 508-15		21
69	Biochemical and genetic exclusion of calmodulin as the site of the basic defect in cystic fibrosis. *Human Genetics*, **1987**, 76, 278-82	6.3	21
68	The Huntington's disease-related cardiomyopathy prevents a hypertrophic response in the R6/2 mouse model. *PLoS ONE*, **2014**, 9, e108961	3.7	21
67	SIRT1 Activity Is Linked to Its Brain Region-Specific Phosphorylation and Is Impaired in Huntington's Disease Mice. *PLoS ONE*, **2016**, 11, e0145425	3.7	21
66	Fragments of HdhQ150 mutant huntingtin form a soluble oligomer pool that declines with aggregate deposition upon aging. *PLoS ONE*, **2012**, 7, e44457	3.7	20
65	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	4.9	19
64	Meso scale discovery-based assays for the detection of aggregated huntingtin. *PLoS ONE*, **2019**, 14, e0213521	3.7	19
63	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	5.6	19
62	Aberrant processing of the Fugu HD (FrHD) mRNA in mouse cells and in transgenic mice. *Human Molecular Genetics*, **1997**, 6, 2141-9	5.6	19
61	Mouse models of triplet repeat diseases. *Molecular Biotechnology*, **2006**, 32, 147-58	3	18
60	Radiation hybrid map spanning the Huntington disease gene region of chromosome 4. *Genomics*, **1992**, 13, 1040-6	4.3	18
59	Systematic interaction network filtering identifies CRMP1 as a novel suppressor of huntingtin misfolding and neurotoxicity. *Genome Research*, **2015**, 25, 701-13	9.7	17

58	The new science of ageing. *Philosophical Transactions of the Royal Society B: Biological Sciences*, **2011**, 366, 6-8	5.8	17
57	Subcellular Localization And Formation Of Huntingtin Aggregates Correlates With Symptom Onset And Progression In A Huntington'S Disease Model. *Brain Communications*, **2020**, 2, fcaa066	4.5	16
56	Novel isoforms of heat shock transcription factor 1, HSF1β and HSF1γ regulate chaperone protein gene transcription. *Journal of Biological Chemistry*, **2014**, 289, 19894-906	5.4	16
55	Sequence-tagged sites (STSs) spanning 4p16.3 and the Huntington disease candidate region. *Genomics*, **1992**, 13, 75-80	4.3	16
54	Polyglutamine expansion and Huntington's disease. *Biochemical Society Transactions*, **1998**, 26, 471-5	5.1	15
53	TBK1 phosphorylates mutant Huntingtin and suppresses its aggregation and toxicity in Huntington's disease models. *EMBO Journal*, **2020**, 39, e104671	13	15
52	The isolation of cDNAs within the Huntington disease region by hybridisation of yeast artificial chromosomes to a cDNA library. *Human Molecular Genetics*, **1993**, 2, 305-9	5.6	13
51	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	0.9	12
50	Myostatin inhibition prevents skeletal muscle pathophysiology in Huntington's disease mice. *Scientific Reports*, **2017**, 7, 14275	4.9	11
49	Correlations of Behavioral Deficits with Brain Pathology Assessed through Longitudinal MRI and Histopathology in the HdhQ150/Q150 Mouse Model of Huntington's Disease. *PLoS ONE*, **2017**, 12, e0168556	3.7	11
48	BIOMEDICINE: One Misfolded Protein Allows Others to Sneak By. *Science*, **2006**, 311, 1385-6	33.3	11
47	Disruption to schizophrenia-associated gene Fez1 in the hippocampus of HDAC11 knockout mice. *Scientific Reports*, **2017**, 7, 11900	4.9	10
46	Live-cell super-resolution microscopy reveals a primary role for diffusion in polyglutamine-driven aggresome assembly. *Journal of Biological Chemistry*, **2019**, 294, 257-268	5.4	10
45	MEF2 impairment underlies skeletal muscle atrophy in polyglutamine disease. *Acta Neuropathologica*, **2020**, 140, 63-80	14.3	10
44	In vivo neutralization of the protagonist role of macrophages during the chronic inflammatory stage of Huntington's disease. *Scientific Reports*, **2018**, 8, 11447	4.9	9
43	RNA Related Pathology in Huntington's Disease. *Advances in Experimental Medicine and Biology*, **2018**, 1049, 85-101	3.6	8
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