Gillian P Bates

List of Publications by Year in Descending Order

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	Uninterrupted CAG repeat drives striatum-selective transcriptionopathy and nuclear pathogenesis in human Huntingtin BAC mice.. *Neuron*, **2022**,	13.9	2
218	Ablation of kynurenine 3-monooxygenase rescues plasma inflammatory cytokine levels in the R6/2 mouse model of Huntington's disease. *Scientific Reports*, **2021**, 11, 5484	4.9	3
217	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	4.9	2
216	Use of high-content imaging to quantify transduction of AAV-PHP viruses in the brain following systemic delivery. *Brain Communications*, **2021**, 3, fcab105	4.5	3
215	Correlative light and electron microscopy suggests that mutant huntingtin dysregulates the endolysosomal pathway in presymptomatic Huntington's disease. *Acta Neuropathologica Communications*, **2021**, 9, 70	7.3	1
214	Small, Seeding-Competent Huntingtin Fibrils Are Prominent Aggregate Species in Brains of zQ175 Huntington's Disease Knock-in Mice. *Frontiers in Neuroscience*, **2021**, 15, 682172	5.1	2
213	Development of novel bioassays to detect soluble and aggregated Huntingtin proteins on three technology platforms. *Brain Communications*, **2021**, 3, fcaa231	4.5	5
212	FAN1 controls mismatch repair complex assembly via MLH1 retention to stabilize CAG repeat expansion in Huntington's disease. *Cell Reports*, **2021**, 36, 109649	10.6	8
211	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
210	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
209	TBK1 phosphorylates mutant Huntingtin and suppresses its aggregation and toxicity in Huntington's disease models. *EMBO Journal*, **2020**, 39, e104671	13	15
208	Silencing Srsf6 does not modulate incomplete splicing of the huntingtin gene in Huntington's disease models. *Scientific Reports*, **2020**, 10, 14057	4.9	6
207	MEF2 impairment underlies skeletal muscle atrophy in polyglutamine disease. *Acta Neuropathologica*, **2020**, 140, 63-80	14.3	10
206	Inhibition of tumour necrosis factor alpha in the R6/2 mouse model of Huntington's disease by etanercept treatment. *Scientific Reports*, **2019**, 9, 7202	4.9	7
205	Meso scale discovery-based assays for the detection of aggregated huntingtin. *PLoS ONE*, **2019**, 14, e0213521	3.7	19
204	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
203	Genetic deletion of S6k1 does not rescue the phenotypic deficits observed in the R6/2 mouse model of Huntington's disease. *Scientific Reports*, **2019**, 9, 16133	4.9	1

202	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	4.9	3
201	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
200	RNA Related Pathology in Huntington's Disease. *Advances in Experimental Medicine and Biology*, **2018**, 1049, 85-101	3.6	8
199	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
198	Regulatory mechanisms of incomplete huntingtin mRNA splicing. *Nature Communications*, **2018**, 9, 3955	17.4	27
197	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
196	Mouse Models of Huntington's Disease. *Methods in Molecular Biology*, **2018**, 1780, 97-120	1.4	34
195	The pathogenic exon 1 HTT protein is produced by incomplete splicing in Huntington's disease patients. *Scientific Reports*, **2017**, 7, 1307	4.9	89
194	Myostatin inhibition prevents skeletal muscle pathophysiology in Huntington's disease mice. *Scientific Reports*, **2017**, 7, 14275	4.9	11
193	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
192	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
191	Disruption to schizophrenia-associated gene Fez1 in the hippocampus of HDAC11 knockout mice. *Scientific Reports*, **2017**, 7, 11900	4.9	10
190	Stall in Canonical Autophagy-Lysosome Pathways Prompts Nucleophagy-Based Nuclear Breakdown in Neurodegeneration. *Current Biology*, **2017**, 27, 3626-3642.e6	6.3	33
189	Frequency of nuclear mutant huntingtin inclusion formation in neurons and glia is cell-type-specific. *Glia*, **2017**, 65, 50-61	9	56
188	L3 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	5.5	
187	B10 Inclusion formation in mutant HTT exon 1 expressing human neuronal cells. *Journal of Neurology, Neurosurgery and Psychiatry*, **2016**, 87, A12.2-A12	5.5	
186	B38 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	5.5	3
185	B24 Assessment of immune system activation status during the course of disease in huntington disease mouse model. *Journal of Neurology, Neurosurgery and Psychiatry*, **2016**, 87, A17.2-A17	5.5	

184	B8 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	5.5	
183	B4 Detection of the aberrantly spliced exon 1 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	5.5	
182	B27 Abnormal bioenergetics in inclusion-containing mutant HTT exon 1 primary human neurons. *Journal of Neurology, Neurosurgery and Psychiatry*, **2016**, 87, A18.2-A19	5.5	
181	B3 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	5.5	
180	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
179	Embryonic Mutant Huntingtin Aggregate Formation in Mouse Models of Huntington's Disease. *Journal of Huntingtons Disease*, **2016**, 5, 343-346	1.9	7
178	B6 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	5.5	
177	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
176	UBQLN2 Mediates Autophagy-Independent Protein Aggregate Clearance by the Proteasome. *Cell*, **2016**, 166, 935-949	56.2	186
175	HDAC4-myogenin axis as an important marker of HD-related skeletal muscle atrophy. *PLoS Genetics* , **2015**, 11, e1005021	6	44
174	Systematic interaction network filtering identifies CRMP1 as a novel suppressor of huntingtin misfolding and neurotoxicity. *Genome Research*, **2015**, 25, 701-13	9.7	17
173	Huntington disease. *Nature Reviews Disease Primers*, **2015**, 1, 15005	51.1	672
172	Treating the whole body in Huntington's disease. *Lancet Neurology, The*, **2015**, 14, 1135-42	24.1	96
171	Alan Walker (ed.), The New Science of Ageing, Policy Press, Bristol, UK, 2014, 344 pp., pbk £26.99, ISBN 13: 9781447314677.. *Ageing and Society*, **2015**, 35, 1796-1797	1.7	
170	In Vivo Profiling Reveals a Competent Heat Shock Response in Adult Neurons: Implications for Neurodegenerative Disorders. *PLoS ONE*, **2015**, 10, e0131985	3.7	7
169	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
168	Characterization of Gastric Mucosa Biopsies Reveals Alterations in Huntington's Disease. *PLOS Currents*, **2015**, 7,		5
167	HTT-lowering reverses Huntington's disease immune dysfunction caused by NFB pathway dysregulation. *Brain*, **2014**, 137, 819-33	11.2	109

166	Dynamic recruitment of active proteasomes into polyglutamine initiated inclusion bodies. *FEBS Letters*, **2014**, 588, 151-9	3.8	40
165	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
164	Dysfunction of the CNS-heart axis in mouse models of Huntington's disease. *PLoS Genetics*, **2014**, 10, e1004550	6	65
163	A common gene expression signature in Huntington's disease patient brain regions. *BMC Medical Genomics*, **2014**, 7, 60	3.7	34
162	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
161	The PDE1/5 Inhibitor SCH-51866 Does Not Modify Disease Progression in the R6/2 Mouse Model of Huntington's Disease. *PLOS Currents*, **2014**, 6,		3
160	Reducing Igf-1r levels leads to paradoxical and sexually dimorphic effects in HD mice. *PLoS ONE*, **2014**, 9, e105595	3.7	6
159	The Huntington's disease-related cardiomyopathy prevents a hypertrophic response in the R6/2 mouse model. *PLoS ONE*, **2014**, 9, e108961	3.7	21
158	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
157	SUMO-2 and PIAS1 modulate insoluble mutant huntingtin protein accumulation. *Cell Reports*, **2013**, 4, 362-75	10.6	68
156	HDAC4 reduction: a novel therapeutic strategy to target cytoplasmic huntingtin and ameliorate neurodegeneration. *PLoS Biology*, **2013**, 11, e1001717	9.7	117
155	Aberrantly spliced HTT, a new player in Huntington's disease pathogenesis. *RNA Biology*, **2013**, 10, 1647-53	4.8	40
154	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
153	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
152	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
151	HDAC4 does not act as a protein deacetylase in the postnatal murine brain in vivo. *PLoS ONE*, **2013**, 8, e80849	3.7	27
150	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
149	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

148	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
147	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
146	Suppression of protein aggregation by chaperone modification of high molecular weight complexes. *Brain*, **2012**, 135, 1180-96	11.2	85
145	Mutant huntingtin fragmentation in immune cells tracks Huntington's disease progression. *Journal of Clinical Investigation*, **2012**, 122, 3731-6	15.9	97
144	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
143	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
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	3.7	31
141	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
140	Gastrointestinal dysfunction contributes to weight loss in Huntington's disease mice. *Neurobiology of Disease*, **2011**, 44, 1-8	7.5	60
139	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
138	The new science of ageing. *Philosophical Transactions of the Royal Society B: Biological Sciences*, **2011**, 366, 6-8	5.8	17
137	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
136	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
135	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
134	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
133	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
132	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
131	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

130	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
129	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
128	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
127	IKK phosphorylates Huntingtin and targets it for degradation by the proteasome and lysosome. *Journal of Cell Biology*, **2009**, 187, 1083-99	7.3	287
126	Systematic behavioral evaluation of Huntington's disease transgenic and knock-in mouse models. *Neurobiology of Disease*, **2009**, 35, 319-36	7.5	244
125	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
124	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
123	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
122	Increased metabolism in the R6/2 mouse model of Huntington's disease. *Neurobiology of Disease*, **2008**, 29, 41-51	7.5	105
121	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
120	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
119	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
118	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
117	Global changes to the ubiquitin system in Huntington's disease. *Nature*, **2007**, 448, 704-8	50.4	444
116	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
115	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
114	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
113	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

112	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
111	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
110	Elevated brain 3-hydroxykynurenine and quinolinate levels in Huntington disease mice. *Neurobiology of Disease*, **2006**, 23, 190-7	7.5	125
109	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
108	BIOMEDICINE: One Misfolded Protein Allows Others to Sneak By. *Science*, **2006**, 311, 1385-6	33.3	11
107	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
106	Molecular Pathogenesis and Therapeutic Targets in Huntington's Disease **2006**, 223-249		1
105	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
104	Histone deacetylase inhibitors as therapeutics for polyglutamine disorders. *Nature Reviews Neuroscience*, **2006**, 7, 784-96	13.5	176
103	Mouse models of triplet repeat diseases. *Molecular Biotechnology*, **2006**, 32, 147-58	3	18
102	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
101	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
100	Biomarkers for neurodegenerative diseases. *Current Opinion in Neurology*, **2005**, 18, 698-705	7.1	55
99	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
98	Polyglutamine expansion of huntingtin impairs its nuclear export. *Nature Genetics*, **2005**, 37, 198-204	36.3	129
97	History of genetic disease: the molecular genetics of Huntington disease - a history. *Nature Reviews Genetics*, **2005**, 6, 766-73	30.1	132
96	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
95	Dysfunction of the cholesterol biosynthetic pathway in Huntington's disease. *Journal of Neuroscience*, **2005**, 25, 9932-9	6.6	195

94	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
93	Mouse models of triplet repeat diseases. *Methods in Molecular Biology*, **2004**, 277, 3-15	1.4	1
92	Monitoring aggregate formation in organotypic slice cultures from transgenic mice. *Methods in Molecular Biology*, **2004**, 277, 161-71	1.4	3
91	Huntingtin and the molecular pathogenesis of Huntington's disease. Fourth in molecular medicine review series. *EMBO Reports*, **2004**, 5, 958-63	6.5	365
90	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
89	Experimental therapeutics in Huntington's disease. *Current Opinion in Neurology*, **2003**, 16, 465-470	7.1	6
88	Experimental therapeutics in Huntington's disease: are models useful for therapeutic trials?. *Current Opinion in Neurology*, **2003**, 16, 465-70	7.1	37
87	Minocycline and doxycycline are not beneficial in a model of Huntington's disease. *Annals of Neurology*, **2003**, 54, 186-96	9.4	145
86	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
85	Inclusion formation in Huntington's disease R6/2 mouse muscle cultures. *Journal of Neurochemistry*, **2003**, 87, 1-6	6	35
84	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
83	Huntingtin aggregation and toxicity in Huntington's disease. *Lancet, The*, **2003**, 361, 1642-4	40	422
82	Standardization and statistical approaches to therapeutic trials in the R6/2 mouse. *Brain Research Bulletin*, **2003**, 61, 469-79	3.9	125
81	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
80	Environmental enrichment slows disease progression in R6/2 Huntington's disease mice. *Annals of Neurology*, **2002**, 51, 235-42	9.4	282
79	Arfaptin 2 regulates the aggregation of mutant huntingtin protein. *Nature Cell Biology*, **2002**, 4, 240-5	23.4	43
78	Alterations in the mouse and human proteome caused by Huntington's disease. *Molecular and Cellular Proteomics*, **2002**, 1, 366-75	7.6	66
77	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

76	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
75	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
74	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
73	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
72	Impaired glutamate uptake in the R6 Huntington's disease transgenic mice. *Neurobiology of Disease* , **2001**, 8, 807-21	7.5	233
71	Inhibition of polyglutamine aggregation in R6/2 HD brain slices-complex dose-response profiles. *Neurobiology of Disease*, **2001**, 8, 1017-26	7.5	66
70	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
69	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
68	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
67	Amyloid-like inclusions in Huntington's disease. *Neuroscience*, **2000**, 100, 677-80	3.9	83
66	Transgenic Mouse Models of Huntington Disease **2000**, 355-367		1
65	Mitochondrial dysfunction and free radical damage in the Huntington R6/2 transgenic mouse. *Annals of Neurology*, **2000**, 47, 80-6	9.4	114
64	Brain neurotransmitter deficits in mice transgenic for the Huntington's disease mutation. *Journal of Neurochemistry*, **1999**, 72, 1773-6	6	72
63	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
62	Selective discrimination learning impairments in mice expressing the human Huntington's disease mutation. *Journal of Neuroscience*, **1999**, 19, 10428-37	6.6	330
61	Formation of polyglutamine inclusions in non-CNS tissue. *Human Molecular Genetics*, **1999**, 8, 813-22	5.6	243
60	Ultrastructural localization and progressive formation of neuropil aggregates in Huntington's disease transgenic mice. *Human Molecular Genetics*, **1999**, 8, 1227-36	5.6	152
59	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

58	Transgenic models of Huntington's disease. *Philosophical Transactions of the Royal Society B: Biological Sciences*, **1999**, 354, 963-9	5.8	58
57	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
56	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
55	Detection of polyglutamine aggregation in mouse models. *Methods in Enzymology*, **1999**, 309, 687-701	1.7	26
54	Molecular Pathology of Huntington's Disease: Animal Models and Nuclear Mechanisms. *Neuroscientist*, **1999**, 5, 383-391	7.6	3
53	Transgenic mice in the study of polyglutamine repeat expansion diseases. *Brain Pathology*, **1998**, 8, 699-714	7.6	49
52	A novel gene encoding an integral membrane protein is mutated in nephropathic cystinosis. *Nature Genetics*, **1998**, 18, 319-24	36.3	458
51	Striking changes in anxiety in Huntington's disease transgenic mice. *Brain Research*, **1998**, 805, 234-40	3.7	61
50	Are neuronal intranuclear inclusions the common neuropathology of triplet-repeat disorders with polyglutamine-repeat expansions?. *Lancet, The*, **1998**, 351, 131-3	40	163
49	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
48	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
47	Striatal transplantation in a transgenic mouse model of Huntington's disease. *Experimental Neurology*, **1998**, 154, 31-40	5.7	105
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