Michael R Hayden

List of Publications by Year
in descending order

765 papers	72,901 citations	435 131 h-index	1222 227 g-index
793 all docs	793 docs citations	793 times ranked	41418 citing authors

#	Article	IF	Citations
1	Mutations in ABC1 in Tangier disease and familial high-density lipoprotein deficiency. Nature Genetics, 1999, 22, 336-345.	21.4	1,609
2	Loss of Huntingtin-Mediated BDNF Gene Transcription in Huntington's Disease. Science, 2001, 293, 493-498.	12.6	1,191
3	Huntington disease. Nature Reviews Disease Primers, 2015, 1, 15005.	30.5	1,031
4	The relationship between trinucleotide (CAG) repeat length and clinical features of Huntington's disease. Nature Genetics, 1993, 4, 398-403.	21.4	1,002
5	Early mitochondrial calcium defects in Huntington's disease are a direct effect of polyglutamines. Nature Neuroscience, 2002, 5, 731-736.	14.8	925
6	Mutations in HFE2 cause iron overload in chromosome 1qâ€“linked juvenile hemochromatosis. Nature Genetics, 2004, 36, 77-82.	21.4	900
7	Huntingtin interacts with REST/NRSF to modulate the transcription of NRSE-controlled neuronal genes. Nature Genetics, 2003, 35, 76-83.	21.4	807
8	A YAC Mouse Model for Huntingtonâ€™s Disease with Full-Length Mutant Huntingtin, Cytoplasmic Toxicity, and Selective Striatal Neurodegeneration. Neuron, 1999, 23, 181-192.	8.1	789
9	Targeted disruption of the Huntington's disease gene results in embryonic lethality and behavioral and morphological changes in heterozygotes. Cell, 1995, 81, 811-823.	28.9	758
10	A new model for prediction of the age of onset and penetrance for Huntington's disease based on CAG length. Clinical Genetics, 2004, 65, 267-277.	2.0	738
11	Selective striatal neuronal loss in a YAC128 mouse model of Huntington disease. Human Molecular Genetics, 2003, 12, 1555-1567.	2.9	713
12	Detection of Huntington's disease decades before diagnosis: the Predict-HD study. Journal of Neurology, Neurosurgery and Psychiatry, 2008, 79, 874-880.	1.9	696
13	A gene encoding a putative GTPase regulator is mutated in familial amyotrophic lateral sclerosis 2. Nature Genetics, 2001, 29, 166-173.	21.4	635
14	Cleavage at the Caspase-6 Site Is Required for Neuronal Dysfunction and Degeneration Due to Mutant Huntingtin. Cell, 2006, 125, 1179-1191.	28.9	600
15	A Worldwide Study of the Huntington's Disease Mutation: The Sensitivity and Specificity of Measuring CAG Repeats. New England Journal of Medicine, 1994, 330, 1401-1406.	27.0	563
16	A novel pathogenic pathway of immune activation detectable before clinical onset in Huntington's disease. Journal of Experimental Medicine, 2008, 205, 1869-1877.	8.5	559
17	Increased Sensitivity to N-Methyl-D-Aspartate Receptor-Mediated Excitotoxicity in a Mouse Model of Huntington's Disease. Neuron, 2002, 33, 849-860.	8.1	553
18	Cleavage of huntingtin by apopain, a proapoptotic cysteine protease, is modulated by the polyglutamine tract. Nature Genetics, 1996, 13, 442-449.	21.4	545

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19	Caspase Cleavage of Gene Products Associated with Triplet Expansion Disorders Generates Truncated Fragments Containing the Polyglutamine Tract. Journal of Biological Chemistry, 1998, 273, 9158-9167.	3.4	499
20	Mutant huntingtin binds the mitochondrial fission GTPase dynamin-related protein-1 and increases its enzymatic activity. Nature Medicine, 2011, 17, 377-382.	30.7	467
21	Length of huntingtin and its polyglutamine tract influences localization and frequency of intracellular aggregates. Nature Genetics, 1998, 18, 150-154.	21.4	456
22	Mutant Huntingtin Impairs Axonal Trafficking in Mammalian Neurons In Vivo and In Vitro. Molecular and Cellular Biology, 2004, 24, 8195-8209.	2.3	454
23	Early Increase in Extrasynaptic NMDA Receptor Signaling and Expression Contributes to Phenotype Onset in Huntington's Disease Mice. Neuron, 2010, 65, 178-190.	8.1	448
24	The Psychological Consequences of Predictive Testing for Huntingtonâ€™s Disease. New England Journal of Medicine, 1992, 327, 1401-1405.	27.0	447
25	Intestinal ABCA1 directly contributes to HDL biogenesis in vivo. Journal of Clinical Investigation, 2006, 116, 1052-1062.	8.2	447
26	Huntingtin and Huntingtin-Associated Protein 1 Influence Neuronal Calcium Signaling Mediated by Inositol-(1,4,5) Triphosphate Receptor Type 1. Neuron, 2003, 39, 227-239.	8.1	442
27	Mutant frizzled-4 disrupts retinal angiogenesis in familial exudative vitreoretinopathy. Nature Genetics, 2002, 32, 326-330.	21.4	409
28	Targeted inactivation of hepatic Abca1 causes profound hypoalphalipoproteinemia and kidney hypercatabolism of apoA-I. Journal of Clinical Investigation, 2005, 115, 1333-1342.	8.2	407
29	Lossâ€ofâ€function mutations in the Na_v1.7 gene underlie congenital indifference to pain in multiple human populations. Clinical Genetics, 2007, 71, 311-319.	2.0	404
30	Differential modulation of endotoxin responsiveness by human caspase-12 polymorphisms. Nature, 2004, 429, 75-79.	27.8	395
31	Somatic and gonadal mosaicism of the Huntington disease gene CAG repeat in brain and sperm. Nature Genetics, 1994, 6, 409-414.	21.4	383
32	Balance between synaptic versus extrasynaptic NMDA receptor activity influences inclusions and neurotoxicity of mutant huntingtin. Nature Medicine, 2009, 15, 1407-1413.	30.7	381
33	Î²-cell ABCA1 influences insulin secretion, glucose homeostasis and response to thiazolidinedione treatment. Nature Medicine, 2007, 13, 340-347.	30.7	366
34	HIP1, a human homologue of S. cerevisiae Sla2p, interacts with membrane-associated huntingtin in the brain. Nature Genetics, 1997, 16, 44-53.	21.4	353
35	Wild-Type Huntingtin Protects from Apoptosis Upstream of Caspase-3. Journal of Neuroscience, 2000, 20, 3705-3713.	3.6	349
36	Mutations in the ABC 1 gene in familial HDL deficiency with defective cholesterol efflux. Lancet, The, 1999, 354, 1341-1346.	13.7	345

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37	Caspase Cleavage of Mutant Huntingtin Precedes Neurodegeneration in Huntington's Disease. Journal of Neuroscience, 2002, 22, 7862-7872.	3.6	344
38	IKK phosphorylates Huntingtin and targets it for degradation by the proteasome and lysosome. Journal of Cell Biology, 2009, 187, 1083-1099.	5.2	343
39	Pharmacogenomic Prediction of Anthracycline-Induced Cardiotoxicity in Children. Journal of Clinical Oncology, 2012, 30, 1422-1428.	1.6	341
40	Disturbed Ca2+ signaling and apoptosis of medium spiny neurons in Huntington's disease. Proceedings of the National Academy of Sciences of the United States of America, 2005, 102, 2602-2607.	7.1	336
41	More Codeine Fatalities After Tonsillectomy in North American Children. Pediatrics, 2012, 129, e1343-e1347.	2.1	334
42	Deficiency of ABCA1 Impairs Apolipoprotein E Metabolism in Brain. Journal of Biological Chemistry, 2004, 279, 41197-41207.	3.4	321
43	Inhibiting Caspase Cleavage of Huntingtin Reduces Toxicity and Aggregate Formation in Neuronal and Nonneuronal Cells. Journal of Biological Chemistry, 2000, 275, 19831-19838.	3.4	320
44	Relationship between stearoyl-CoA desaturase activity and plasma triglycerides in human and mouse hypertriglyceridemia. Journal of Lipid Research, 2002, 43, 1899-1907.	4.2	318
45	Huntingtin Is Ubiquitinated and Interacts with a Specific Ubiquitin-conjugating Enzyme. Journal of Biological Chemistry, 1996, 271, 19385-19394.	3.4	313
46	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.	2.9	304
47	CAG repeat expansion in Huntington disease determines age at onset in a fully dominant fashion. Neurology, 2012, 78, 690-695.	1.1	303
48	The Influence of Huntingtin Protein Size on Nuclear Localization and Cellular Toxicity. Journal of Cell Biology, 1998, 141, 1097-1105.	5.2	300
49	Age and residual cholesterol efflux affect HDL cholesterol levels and coronary artery disease in ABCA1 heterozygotes. Journal of Clinical Investigation, 2000, 106, 1263-1270.	8.2	295
50	A one-hit model of cell death in inherited neuronal degenerations. Nature, 2000, 406, 195-199.	27.8	294
51	Cell death attenuation by `Usurpin', a mammalian DED-caspase homologue that precludes caspase-8 recruitment and activation by the CD-95 (Fas, APO-1) receptor complex. Cell Death and Differentiation, 1998, 5, 271-288.	11.2	293
52	Preparing for Preventive Clinical Trials. Archives of Neurology, 2006, 63, 883.	4.5	292
53	CAGâ€repeat length and the age of onset in Huntington disease (HD): A review and validation study of statistical approaches. American Journal of Medical Genetics Part B: Neuropsychiatric Genetics, 2010, 153B, 397-408.	1.7	289
54	Genetic variants in TPMT and COMT are associated with hearing loss in children receiving cisplatin chemotherapy. Nature Genetics, 2009, 41, 1345-1349.	21.4	287

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55	Choosing an animal model for the study of Huntington's disease. Nature Reviews Neuroscience, 2013, 14, 708-721.	10.2	287
56	Premature Atherosclerosis in Patients with Familial Chylomicronemia Caused by Mutations in the Lipoprotein Lipase Gene. New England Journal of Medicine, 1996, 335, 848-854.	27.0	286
57	Pharmacogenetics of Neonatal Opioid Toxicity Following Maternal Use of Codeine During Breastfeeding: A Caseâ€"Control Study. Clinical Pharmacology and Therapeutics, 2009, 85, 31-35.	4.7	284
58	Restoration of Endothelial Function by Increasing High-Density Lipoprotein in Subjects With Isolated Low High-Density Lipoprotein. Circulation, 2003, 107, 2944-2948.	1.6	283
59	Cognitive Dysfunction Precedes Neuropathology and Motor Abnormalities in the YAC128 Mouse Model of Huntington's Disease. Journal of Neuroscience, 2005, 25, 4169-4180.	3.6	282
60	Common Genetic Variation inABCA1Is Associated With Altered Lipoprotein Levels and a Modified Risk for Coronary Artery Disease. Circulation, 2001, 103, 1198-1205.	1.6	280
61	Autophagy in Huntington disease and huntingtin in autophagy. Trends in Neurosciences, 2015, 38, 26-35.	8.6	277
62	Recruitment and activation of caspase-8 by the Huntingtin-interacting protein Hip-1 and a novel partner Hippi. Nature Cell Biology, 2002, 4, 95-105.	10.3	274
63	A Worldwide Assessment of the Frequency of Suicide, Suicide Attempts, or Psychiatric Hospitalization after Predictive Testing for Huntington Disease. American Journal of Human Genetics, 1999, 64, 1293-1304.	6.2	271
64	Huntingtin-Interacting Protein HIP14 Is a Palmitoyl Transferase Involved in Palmitoylation and Trafficking of Multiple Neuronal Proteins. Neuron, 2004, 44, 977-986.	8.1	271
65	Palmitoylation of huntingtin by HIP14is essential for its trafficking and function. Nature Neuroscience, 2006, 9, 824-831.	14.8	266
66	ABCA1 mRNA and Protein Distribution Patterns Predict Multiple Different Roles and Levels of Regulation. Laboratory Investigation, 2002, 82, 273-283.	3.7	265
67	Molecular analysis of new mutations for Huntington's disease: intermediate alleles and sex of origin effects. Nature Genetics, 1993, 5, 174-179.	21.4	248
68	Absence of behavioral abnormalities and neurodegeneration in vivo despite widespread neuronal huntingtin inclusions. Proceedings of the National Academy of Sciences of the United States of America, 2005, 102, 11402-11407.	7.1	247
69	NMDA receptor function in mouse models of Huntington disease. Journal of Neuroscience Research, 2001, 66, 525-539.	2.9	246
70	When good drugs go bad. Nature, 2007, 446, 975-977.	27.8	246
71	Potent and Selective Antisense Oligonucleotides Targeting Single-Nucleotide Polymorphisms in the Huntington Disease Gene / Allele-Specific Silencing of Mutant Huntingtin. Molecular Therapy, 2011, 19, 2178-2185.	8.2	246
72	Huntingtonâ€™s Chorea. , 1981, , .		246

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73	Pivotal role of ABCA1 in reverse cholesterol transport influencing HDL levels and susceptibility to atherosclerosis. Journal of Lipid Research, 2001, 42, 1717-1726.	4.2	244
74	Inhibition of Calpain Cleavage of Huntingtin Reduces Toxicity. Journal of Biological Chemistry, 2004, 279, 20211-20220.	3.4	242
75	Deranged neuronal calcium signaling and Huntington disease. Biochemical and Biophysical Research Communications, 2004, 322, 1310-1317.	2.1	236
76	A lipoprotein lipase mutation (Asn291Ser) is associated with reduced HDL cholesterol levels in premature atherosclerosis. Nature Genetics, 1995, 10, 28-34.	21.4	235
77	Efflux and Atherosclerosis. Arteriosclerosis, Thrombosis, and Vascular Biology, 2003, 23, 1322-1332.	2.4	231
78	Mutant DNA-binding domain of HSF4 is associated with autosomal dominant lamellar and Marner cataract. Nature Genetics, 2002, 31, 276-278.	21.4	229
79	Predictive testing for Huntington disease in Canada: Adverse effects and unexpected results in those receiving a decreased risk. American Journal of Medical Genetics Part A, 1992, 42, 508-515.	2.4	225
80	Targeted inactivation of hepatic Abca1 causes profound hypoalphalipoproteinemia and kidney hypercatabolism of apoA-I. Journal of Clinical Investigation, 2005, 115, 1333-1342.	8.2	225
81	BDNF Overexpression in the Forebrain Rescues Huntington's Disease Phenotypes in YAC128 Mice. Journal of Neuroscience, 2010, 30, 14708-14718.	3.6	223
82	Increased ABCA1 activity protects against atherosclerosis. Journal of Clinical Investigation, 2002, 110, 35-42.	8.2	216
83	A coding variant in RARG confers susceptibility to anthracycline-induced cardiotoxicity in childhood cancer. Nature Genetics, 2015, 47, 1079-1084.	21.4	214
84	Kennedy's Disease. Journal of Neurochemistry, 1999, 72, 185-195.	3.9	211
85	Wild-Type Huntingtin Reduces the Cellular Toxicity of Mutant Huntingtin In Vivo. American Journal of Human Genetics, 2001, 68, 313-324.	6.2	210
86	CAG Expansion in the Huntington Disease Gene Is Associated with a Specific and Targetable Predisposing Haplogroup. American Journal of Human Genetics, 2009, 84, 351-366.	6.2	204
87	The Absence of ABCA1 Decreases Soluble ApoE Levels but Does Not Diminish Amyloid Deposition in Two Murine Models of Alzheimer Disease. Journal of Biological Chemistry, 2005, 280, 43243-43256.	3.4	203
88	An Actin-Binding Protein of the Sla2/Huntingtin Interacting Protein 1 Family Is a Novel Component of Clathrin-Coated Pits and Vesicles. Journal of Cell Biology, 1999, 147, 1503-1518.	5.2	202
89	Intramuscular Administration of AAV1-Lipoprotein Lipase ^{S447X} Lowers Triglycerides in Lipoprotein Lipaseâ€"Deficient Patients. Arteriosclerosis, Thrombosis, and Vascular Biology, 2008, 28, 2303-2304.	2.4	201
90	Expanded polyglutamines in Caenorhabditis elegans cause axonal abnormalities and severe dysfunction of PLM mechanosensory neurons without cell death. Proceedings of the National Academy of Sciences of the United States of America, 2001, 98, 13318-13323.	7.1	199

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91	Clinical markers of early disease in persons near onset of Huntingtonâ€™s disease. Neurology, 2001, 57, 658-662.	1.1	197
92	Specific caspase interactions and amplification are involved in selective neuronal vulnerability in Huntington's disease. Cell Death and Differentiation, 2004, 11, 424-438.	11.2	193
93	Molecular analysis of juvenile Huntington disease: the major influence on (CAG)n repeat length is the sex of the affected parent. Human Molecular Genetics, 1993, 2, 1535-1540.	2.9	189
94	HIP14, a novel ankyrin domain-containing protein, links huntingtin to intracellular trafficking and endocytosis. Human Molecular Genetics, 2002, 11, 2815-2828.	2.9	189
95	Recommendations for genetic testing to reduce the incidence of anthracyclineâ€induced cardiotoxicity. British Journal of Clinical Pharmacology, 2016, 82, 683-695.	2.4	188
96	Association between increased arterial-wall thickness and impairment in ABCA1-driven cholesterol efflux: an observational study. Lancet, The, 2002, 359, 37-41.	13.7	186
97	Beyond disgust: impaired recognition of negative emotions prior to diagnosis in Huntington's disease. Brain, 2007, 130, 1732-1744.	7.6	181
98	DNA haplotype analysis of Huntington disease reveals clues to the origins and mechanisms of CAG expansion and reasons for geographic variations of prevalence. Human Molecular Genetics, 1994, 3, 2103-2114.	2.9	178
99	Human ABCA1 BAC Transgenic Mice Show Increased High Density Lipoprotein Cholesterol and ApoAI-dependent Efflux Stimulated by an Internal Promoter Containing Liver X Receptor Response Elements in Intron 1. Journal of Biological Chemistry, 2001, 276, 33969-33979.	3.4	176
100	HIP1 Functions in Clathrin-mediated Endocytosis through Binding to Clathrin and Adaptor Protein 2. Journal of Biological Chemistry, 2001, 276, 39271-39276.	3.4	174
101	Mutant Huntingtin Enhances Excitotoxic Cell Death. Molecular and Cellular Neurosciences, 2001, 17, 41-53.	2.2	173
102	Homozygosity for CAG mutation in Huntington disease is associated with a more severe clinical course. Brain, 2003, 126, 946-955.	7.6	173
103	Positron emission tomography in the early diagnosis of Huntington's disease. Neurology, 1986, 36, 888-888.	1.1	171
104	Subtype-Specific Enhancement of NMDA Receptor Currents by Mutant Huntingtin. Journal of Neurochemistry, 2008, 72, 1890-1898.	3.9	166
105	Validation of variants in SLC28A3 and UGT1A6 as genetic markers predictive of anthracyclineâ€induced cardiotoxicity in children. Pediatric Blood and Cancer, 2013, 60, 1375-1381.	1.5	166
106	Apolipoprotein B gene variants are involved in the determination of serum cholesterol levels: a study in normo- and hypelipidaemic individuals. Atherosclerosis, 1987, 67, 81-89.	0.8	165
107	Predictive testing for Huntington disease in Canada: The experience of those receiving an increased risk. American Journal of Medical Genetics Part A, 1992, 42, 499-507.	2.4	163
108	Macrophage ATP-Binding Cassette Transporter A1 Overexpression Inhibits Atherosclerotic Lesion Progression in Low-Density Lipoprotein Receptor Knockout Mice. Arteriosclerosis, Thrombosis, and Vascular Biology, 2006, 26, 929-934.	2.4	163

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109	Genetic and environmental factors affecting the incidence of coronary artery disease in heterozygous familial hypercholesterolemia.. Arteriosclerosis and Thrombosis: A Journal of Vascular Biology, 1991, 11, 290-297.	3.9	160
110	Age-Dependent Alterations of Corticostriatal Activity in the YAC128 Mouse Model of Huntington Disease. Journal of Neuroscience, 2009, 29, 2414-2427.	3.6	160
111	A highly polymorphic locus very tightly linked to the Huntington's disease gene. Nature, 1988, 332, 734-736.	27.8	159
112	Small Changes, Big Impact. Neuroscientist, 2011, 17, 475-492.	3.5	159
113	Differential effect of the rs4149056 variant in SLCO1B1 on myopathy associated with simvastatin and atorvastatin. Pharmacogenomics Journal, 2012, 12, 233-237.	2.0	158
114	A CCG repeat polymorphism adjacent to the CAG repeat in the Huntington disease gene: implications for diagnostic accuracy and predictive testing. Human Molecular Genetics, 1994, 3, 65-67.	2.9	157
115	Huntingtin Bodies Sequester Vesicle-Associated Proteins by a Polyproline-Dependent Interaction. Journal of Neuroscience, 2004, 24, 269-281.	3.6	157
116	A missense mutation at codon 188 of the human lipoprotein lipase gene is a frequent cause of lipoprotein lipase deficiency in persons of different ancestries.. Journal of Clinical Investigation, 1990, 86, 728-734.	8.2	157
117	Selective degeneration and nuclear localization of mutant huntingtin in the YAC128 mouse model of Huntington disease. Human Molecular Genetics, 2005, 14, 3823-3835.	2.9	152
118	Potentiation of NMDA receptor-mediated excitotoxicity linked with intrinsic apoptotic pathway in YAC transgenic mouse model of Huntington's disease. Molecular and Cellular Neurosciences, 2004, 25, 469-479.	2.2	151
119	Automated deformation analysis in the YAC128 Huntington disease mouse model. NeuroImage, 2008, 39, 32-39.	4.2	150
120	Loss of wild-type huntingtin influences motor dysfunction and survival in the YAC128 mouse model of Huntington disease. Human Molecular Genetics, 2005, 14, 1379-1392.	2.9	149
121	A Genome Scan for Modifiers of Age at Onset in Huntington Disease: The HD MAPS Study. American Journal of Human Genetics, 2003, 73, 682-687.	6.2	148
122	Interaction of normal and expanded CAG repeat sizes influences age at onset of Huntington disease. , 2003, 119A, 279-282.		146
123	Wild-type huntingtin protects neurons from excitotoxicity. Journal of Neurochemistry, 2006, 96, 1121-1129.	3.9	145
124	Ethyl-EPA in Huntington disease: A double-blind, randomized, placebo-controlled trial. Neurology, 2005, 65, 286-292.	1.1	143
125	Predictive testing for Huntington disease: II. Demographic characteristics, life-style patterns, attitudes, and psychosocial assessments of the first fifty-one test candidates. American Journal of Medical Genetics Part A, 1989, 32, 217-224.	2.4	141
126	Lipoprotein Lipase S447X. Arteriosclerosis, Thrombosis, and Vascular Biology, 2006, 26, 1236-1245.	2.4	140

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127	HTT haplotypes contribute to differences in Huntington disease prevalence between Europe and East Asia. European Journal of Human Genetics, 2011, 19, 561-566.	2.8	140
128	Variations on a Gene: Rare and Common Variants inABCA1and Their Impact on HDL Cholesterol Levels and Atherosclerosis. Annual Review of Nutrition, 2006, 26, 105-129.	10.1	139
129	Accumulation of N-terminal mutant huntingtin in mouse and monkey models implicated as a pathogenic mechanism in Huntington's disease. Human Molecular Genetics, 2008, 17, 2738-2751.	2.9	139
130	Neuronal palmitoyl acyl transferases exhibit distinct substrate specificity. FASEB Journal, 2009, 23, 2605-2615.	0.5	138
131	Rational design of antisense oligonucleotides targeting single nucleotide polymorphisms for potent and allele selective suppression of mutant Huntingtin in the CNS. Nucleic Acids Research, 2013, 41, 9634-9650.	14.5	138
132	Elevated brain 3-hydroxykynurenine and quinolinate levels in Huntington disease mice. Neurobiology of Disease, 2006, 23, 190-197.	4.4	137
133	Cholesterol Defect Is Marked across Multiple Rodent Models of Huntington's Disease and Is Manifest in Astrocytes. Journal of Neuroscience, 2010, 30, 10844-10850.	3.6	136
134	Prevention of depressive behaviour in the YAC128 mouse model of Huntington disease by mutation at residue 586 of huntingtin. Brain, 2008, 132, 919-932.	7.6	135
135	Predictive, preâ€natal and diagnostic genetic testing for Huntington's disease: the experience in Canada from 1987 to 2000. Clinical Genetics, 2003, 63, 462-475.	2.0	134
136	Identification of a Novel Gene (HSN2) Causing Hereditary Sensory and Autonomic Neuropathy Type II through the Study of Canadian Genetic Isolates. American Journal of Human Genetics, 2004, 74, 1064-1073.	6.2	133
137	Huntingtin Associates with Acidic Phospholipids at the Plasma Membrane. Journal of Biological Chemistry, 2005, 280, 36464-36473.	3.4	133
138	A functional ABCA1 gene variant is associated with low HDL-cholesterol levels and shows evidence of positive selection in Native Americans. Human Molecular Genetics, 2010, 19, 2877-2885.	2.9	133
139	Rethinking Genotype and Phenotype Correlations in Polyglutamine Expansion Disorders. Human Molecular Genetics, 1997, 6, 2005-2010.	2.9	132
140	Modeling Doxorubicin-Induced Cardiotoxicity in Human Pluripotent Stem Cell Derived-Cardiomyocytes. Scientific Reports, 2016, 6, 25333.	3.3	130
141	Length of Uninterrupted CAG, Independent of Polyglutamine Size, Results in Increased Somatic Instability, Hastening Onset of Huntington Disease. American Journal of Human Genetics, 2019, 104, 1116-1126.	6.2	130
142	Increased instability of intermediate alleles in families with sporadic Huntington disease compared to similar sized intermediate alleles in the general population. Human Molecular Genetics, 1995, 4, 1911-1918.	2.9	129
143	Increased huntingtin protein length reduces the number of polyglutamine-induced gene expression changes in mouse models of Huntington's disease. Human Molecular Genetics, 2002, 11, 1939-1951.	2.9	129
144	Phenotypic Variation in Heterozygous Familial Hypercholesterolemia. Arteriosclerosis, Thrombosis, and Vascular Biology, 1998, 18, 309-315.	2.4	128

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145	Als2 -deficient mice exhibit disturbances in endosome trafficking associated with motor behavioral abnormalities. Proceedings of the National Academy of Sciences of the United States of America, 2006, 103, 9595-9600.	7.1	128
146	Huntingtin phosphorylation on serine 421 is significantly reduced in the striatum and by polyglutamine expansion in vivo. Human Molecular Genetics, 2005, 14, 1569-1577.	2.9	126
147	Multisource ascertainment of Huntington disease in Canada: Prevalence and population at risk. Movement Disorders, 2014, 29, 105-114.	3.9	125
148	Cholesterol in islet dysfunction and type 2 diabetes. Journal of Clinical Investigation, 2008, 118, 403-408.	8.2	125
149	A novel apoA-I mutation (L178P) leads to endothelial dysfunction, increased arterial wall thickness, and premature coronary artery disease. Journal of the American College of Cardiology, 2004, 44, 1429-1435.	2.8	124
150	Mitochondrial-Dependent Ca2+ Handling in Huntington's Disease Striatal Cells: Effect of Histone Deacetylase Inhibitors. Journal of Neuroscience, 2006, 26, 11174-11186.	3.6	124
151	Mutant Huntingtin N-terminal Fragments of Specific Size Mediate Aggregation and Toxicity in Neuronal Cells. Journal of Biological Chemistry, 2009, 284, 10855-10867.	3.4	124
152	Differential Susceptibility to Excitotoxic Stress in YAC128 Mouse Models of Huntington Disease between Initiation and Progression of Disease. Journal of Neuroscience, 2009, 29, 2193-2204.	3.6	123
153	Verbal episodic memory declines prior to diagnosis in Huntington's disease. Neuropsychologia, 2007, 45, 1767-1776.	1.6	122
154	Cholesterol metabolism in Huntington disease. Nature Reviews Neurology, 2011, 7, 561-572.	10.1	122
155	Marked differences in neurochemistry and aggregates despite similar behavioural and neuropathological features of Huntington disease in the full-length BACHD and YAC128 mice. Human Molecular Genetics, 2012, 21, 2219-2232.	2.9	122
156	miR-33a Modulates ABCA1 Expression, Cholesterol Accumulation, and Insulin Secretion in Pancreatic Islets. Diabetes, 2012, 61, 653-658.	0.6	122
157	Increased ABCA1 activity protects against atherosclerosis. Journal of Clinical Investigation, 2002, 110, 35-42.	8.2	121
158	HDL and LDL cholesterol significantly influence Î²-cell function in type 2 diabetes mellitus. Current Opinion in Lipidology, 2010, 21, 178-185.	2.7	120
159	Curation of the Mammalian Palmitoylome Indicates a Pivotal Role for Palmitoylation in Diseases and Disorders of the Nervous System and Cancers. PLoS Computational Biology, 2015, 11, e1004405.	3.2	120
160	Caspases and neurodegeneration: on the cutting edge of new therapeutic approaches. Clinical Genetics, 2000, 57, 1-10.	2.0	119
161	Putting proteins in their place: Palmitoylation in Huntington disease and other neuropsychiatric diseases. Progress in Neurobiology, 2012, 97, 220-238.	5.7	118
162	Therapeutic approaches to Huntington disease: from the bench to the clinic. Nature Reviews Drug Discovery, 2018, 17, 729-750.	46.4	117

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163	Genetic Variant Showing a Positive Interaction With Î²-Blocking Agents With a Beneficial Influence on Lipoprotein Lipase Activity, HDL Cholesterol, and Triglyceride Levels in Coronary Artery Disease Patients. Circulation, 1997, 95, 2628-2635.	1.6	117
164	Contribution of DNA Sequence and CAG Size to Mutation Frequencies of Intermediate Alleles for Huntington Disease: Evidence from Single Sperm Analyses. Human Molecular Genetics, 1997, 6, 301-309.	2.9	116
165	Protein Kinase A Site-specific Phosphorylation Regulates ATP-binding Cassette A1 (ABCA1)-mediated Phospholipid Efflux. Journal of Biological Chemistry, 2002, 277, 41835-41842.	3.4	116
166	Specific Loss of Brain ABCA1 Increases Brain Cholesterol Uptake and Influences Neuronal Structure and Function. Journal of Neuroscience, 2009, 29, 3579-3589.	3.6	116
167	Antisense oligonucleotide therapeutics for inherited neurodegenerative diseases. Trends in Molecular Medicine, 2012, 18, 634-643.	6.7	116
168	Treatment of Nav1.7-mediated pain in inherited erythromelalgia using a novel sodium channel blocker. Pain, 2012, 153, 80-85.	4.2	116
169	Cortical thickness measured from MRI in the YAC128 mouse model of Huntington's disease. NeuroImage, 2008, 41, 243-251.	4.2	115
170	In Vivo Evaluation of Candidate Allele-specific Mutant Huntingtin Gene Silencing Antisense Oligonucleotides. Molecular Therapy, 2014, 22, 2093-2106.	8.2	115
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