Michael R Hayden

List of Publications by Citations

746 papers	61,756 citations	125 h-index	213 g-index
792 ext. papers	67,338 ext. citations	9.1 avg, IF	7.18 L-index

#	Paper	IF	Citations
746	Mutations in ABC1 in Tangier disease and familial high-density lipoprotein deficiency. *Nature Genetics*, **1999**, 22, 336-45	36.3	1468
745	Loss of huntingtin-mediated BDNF gene transcription in Huntington's disease. *Science*, **2001**, 293, 493-8	33.3	1044
744	The relationship between trinucleotide (CAG) repeat length and clinical features of Huntington's disease. *Nature Genetics*, **1993**, 4, 398-403	36.3	868
743	Early mitochondrial calcium defects in Huntington's disease are a direct effect of polyglutamines. *Nature Neuroscience*, **2002**, 5, 731-6	25.5	839
742	Mutations in HFE2 cause iron overload in chromosome 1q-linked juvenile hemochromatosis. *Nature Genetics*, **2004**, 36, 77-82	36.3	797
741	A YAC mouse model for Huntington's disease with full-length mutant huntingtin, cytoplasmic toxicity, and selective striatal neurodegeneration. *Neuron*, **1999**, 23, 181-92	13.9	718
740	Huntingtin interacts with REST/NRSF to modulate the transcription of NRSE-controlled neuronal genes. *Nature Genetics*, **2003**, 35, 76-83	36.3	709
739	Huntington disease. *Nature Reviews Disease Primers*, **2015**, 1, 15005	51.1	672
738	Targeted disruption of the Huntington's disease gene results in embryonic lethality and behavioral and morphological changes in heterozygotes. *Cell*, **1995**, 81, 811-23	56.2	671
737	A new model for prediction of the age of onset and penetrance for Huntington's disease based on CAG length. *Clinical Genetics*, **2004**, 65, 267-77	4	614
736	Selective striatal neuronal loss in a YAC128 mouse model of Huntington disease. *Human Molecular Genetics*, **2003**, 12, 1555-67	5.6	609
735	Detection of Huntington's disease decades before diagnosis: the Predict-HD study. *Journal of Neurology, Neurosurgery and Psychiatry*, **2008**, 79, 874-80	5.5	580
734	A gene encoding a putative GTPase regulator is mutated in familial amyotrophic lateral sclerosis 2. *Nature Genetics*, **2001**, 29, 166-73	36.3	552
733	Cleavage at the caspase-6 site is required for neuronal dysfunction and degeneration due to mutant huntingtin. *Cell*, **2006**, 125, 1179-91	56.2	511
732	Increased sensitivity to N-methyl-D-aspartate receptor-mediated excitotoxicity in a mouse model of Huntington's disease. *Neuron*, **2002**, 33, 849-60	13.9	506
731	Cleavage of huntingtin by apopain, a proapoptotic cysteine protease, is modulated by the polyglutamine tract. *Nature Genetics*, **1996**, 13, 442-9	36.3	501
730	A worldwide study of the Huntington's disease mutation. The sensitivity and specificity of measuring CAG repeats. *New England Journal of Medicine*, **1994**, 330, 1401-6	59.2	485

729	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
728	Caspase cleavage of gene products associated with triplet expansion disorders generates truncated fragments containing the polyglutamine tract. *Journal of Biological Chemistry*, **1998**, 273, 9158-67	5.4	424
727	Length of huntingtin and its polyglutamine tract influences localization and frequency of intracellular aggregates. *Nature Genetics*, **1998**, 18, 150-4	36.3	423
726	Mutant huntingtin impairs axonal trafficking in mammalian neurons in vivo and in vitro. *Molecular and Cellular Biology*, **2004**, 24, 8195-209	4.8	409
725	Mutant huntingtin binds the mitochondrial fission GTPase dynamin-related protein-1 and increases its enzymatic activity. *Nature Medicine*, **2011**, 17, 377-82	50.5	400
724	Huntingtin and huntingtin-associated protein 1 influence neuronal calcium signaling mediated by inositol-(1,4,5) triphosphate receptor type 1. *Neuron*, **2003**, 39, 227-39	13.9	392
723	Intestinal ABCA1 directly contributes to HDL biogenesis in vivo. *Journal of Clinical Investigation*, **2006**, 116, 1052-62	15.9	389
722	Early increase in extrasynaptic NMDA receptor signaling and expression contributes to phenotype onset in Huntington's disease mice. *Neuron*, **2010**, 65, 178-90	13.9	377
721	Targeted inactivation of hepatic Abca1 causes profound hypoalphalipoproteinemia and kidney hypercatabolism of apoA-I. *Journal of Clinical Investigation*, **2005**, 115, 1333-1342	15.9	373
720	The psychological consequences of predictive testing for Huntington's disease. Canadian Collaborative Study of Predictive Testing. *New England Journal of Medicine*, **1992**, 327, 1401-5	59.2	372
719	Mutant frizzled-4 disrupts retinal angiogenesis in familial exudative vitreoretinopathy. *Nature Genetics*, **2002**, 32, 326-30	36.3	368
718	Differential modulation of endotoxin responsiveness by human caspase-12 polymorphisms. *Nature*, **2004**, 429, 75-9	50.4	344
717	Balance between synaptic versus extrasynaptic NMDA receptor activity influences inclusions and neurotoxicity of mutant huntingtin. *Nature Medicine*, **2009**, 15, 1407-13	50.5	343
716	Loss-of-function mutations in the Nav1.7 gene underlie congenital indifference to pain in multiple human populations. *Clinical Genetics*, **2007**, 71, 311-9	4	333
715	Somatic and gonadal mosaicism of the Huntington disease gene CAG repeat in brain and sperm. *Nature Genetics*, **1994**, 6, 409-14	36.3	331
714	HIP1, a human homologue of S. cerevisiae Sla2p, interacts with membrane-associated huntingtin in the brain. *Nature Genetics*, **1997**, 16, 44-53	36.3	327
713	Mutations in the ABC1 gene in familial HDL deficiency with defective cholesterol efflux. *Lancet, The* , **1999**, 354, 1341-6	40	321
712	Beta-cell ABCA1 influences insulin secretion, glucose homeostasis and response to thiazolidinedione treatment. *Nature Medicine*, **2007**, 13, 340-7	50.5	315

711	Wild-type huntingtin protects from apoptosis upstream of caspase-3. *Journal of Neuroscience*, **2000**, 20, 3705-13	6.6	310
710	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, 2602-7	11.5	303
709	Relationship between stearoyl-CoA desaturase activity and plasma triglycerides in human and mouse hypertriglyceridemia. *Journal of Lipid Research*, **2002**, 43, 1899-907	6.3	296
708	Caspase cleavage of mutant huntingtin precedes neurodegeneration in Huntington's disease. *Journal of Neuroscience*, **2002**, 22, 7862-72	6.6	293
707	More codeine fatalities after tonsillectomy in North American children. *Pediatrics*, **2012**, 129, e1343-7	7.4	289
706	IKK phosphorylates Huntingtin and targets it for degradation by the proteasome and lysosome. *Journal of Cell Biology*, **2009**, 187, 1083-99	7.3	287
705	The influence of huntingtin protein size on nuclear localization and cellular toxicity. *Journal of Cell Biology*, **1998**, 141, 1097-105	7.3	280
704	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, 271-88	12.7	279
703	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
702	Pharmacogenomic prediction of anthracycline-induced cardiotoxicity in children. *Journal of Clinical Oncology*, **2012**, 30, 1422-8	2.2	267
701	Age and residual cholesterol efflux affect HDL cholesterol levels and coronary artery disease in ABCA1 heterozygotes. *Journal of Clinical Investigation*, **2000**, 106, 1263-70	15.9	266
700	Restoration of endothelial function by increasing high-density lipoprotein in subjects with isolated low high-density lipoprotein. *Circulation*, **2003**, 107, 2944-8	16.7	264
699	Deficiency of ABCA1 impairs apolipoprotein E metabolism in brain. *Journal of Biological Chemistry*, **2004**, 279, 41197-207	5.4	263
698	Huntingtin is ubiquitinated and interacts with a specific ubiquitin-conjugating enzyme. *Journal of Biological Chemistry*, **1996**, 271, 19385-94	5.4	263
697	Common genetic variation in ABCA1 is associated with altered lipoprotein levels and a modified risk for coronary artery disease. *Circulation*, **2001**, 103, 1198-205	16.7	262
696	Preparing for preventive clinical trials: the Predict-HD study. *Archives of Neurology*, **2006**, 63, 883-90		259
695	Premature atherosclerosis in patients with familial chylomicronemia caused by mutations in the lipoprotein lipase gene. *New England Journal of Medicine*, **1996**, 335, 848-54	59.2	256
694	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	23.4	253

693	Inhibiting caspase cleavage of huntingtin reduces toxicity and aggregate formation in neuronal and nonneuronal cells. *Journal of Biological Chemistry*, **2000**, 275, 19831-8	5.4	251
692	A one-hit model of cell death in inherited neuronal degenerations. *Nature*, **2000**, 406, 195-9	50.4	250
691	Cognitive dysfunction precedes neuropathology and motor abnormalities in the YAC128 mouse model of Huntington's disease. *Journal of Neuroscience*, **2005**, 25, 4169-80	6.6	247
690	Huntingtin-interacting protein HIP14 is a palmitoyl transferase involved in palmitoylation and trafficking of multiple neuronal proteins. *Neuron*, **2004**, 44, 977-86	13.9	247
689	Genetic variants in TPMT and COMT are associated with hearing loss in children receiving cisplatin chemotherapy. *Nature Genetics*, **2009**, 41, 1345-9	36.3	243
688	Pharmacogenetics of neonatal opioid toxicity following maternal use of codeine during breastfeeding: a case-control study. *Clinical Pharmacology and Therapeutics*, **2009**, 85, 31-5	6.1	240
687	ABCA1 mRNA and protein distribution patterns predict multiple different roles and levels of regulation. *Laboratory Investigation*, **2002**, 82, 273-83	5.9	239
686	CAG repeat expansion in Huntington disease determines age at onset in a fully dominant fashion. *Neurology*, **2012**, 78, 690-5	6.5	231
685	Palmitoylation of huntingtin by HIP14 is essential for its trafficking and function. *Nature Neuroscience*, **2006**, 9, 824-31	25.5	230
684	Huntington Chorea **1981**,		228
683	Choosing an animal model for the study of Huntington's disease. *Nature Reviews Neuroscience*, **2013**, 14, 708-21	13.5	227
682	CAG-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	3.5	225
681	Molecular analysis of new mutations for Huntington's disease: intermediate alleles and sex of origin effects. *Nature Genetics*, **1993**, 5, 174-9	36.3	224
680	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, 11402-7	11.5	223
679	NMDA receptor function in mouse models of Huntington disease. *Journal of Neuroscience Research*, **2001**, 66, 525-39	4.4	218
678	Mutant DNA-binding domain of HSF4 is associated with autosomal dominant lamellar and Marner cataract. *Nature Genetics*, **2002**, 31, 276-8	36.3	215
677	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, 1293-304	11	214
676	Inhibition of calpain cleavage of huntingtin reduces toxicity: accumulation of calpain/caspase fragments in the nucleus. *Journal of Biological Chemistry*, **2004**, 279, 20211-20	5.4	213

675	A lipoprotein lipase mutation (Asn291Ser) is associated with reduced HDL cholesterol levels in premature atherosclerosis. *Nature Genetics*, **1995**, 10, 28-34	36.3	213
674	Pivotal role of ABCA1 in reverse cholesterol transport influencing HDL levels and susceptibility to atherosclerosis. *Journal of Lipid Research*, **2001**, 42, 1717-1726	6.3	213
673	Efflux and atherosclerosis: the clinical and biochemical impact of variations in the ABCA1 gene. *Arteriosclerosis, Thrombosis, and Vascular Biology*, **2003**, 23, 1322-32	9.4	212
672	Deranged neuronal calcium signaling and Huntington disease. *Biochemical and Biophysical Research Communications*, **2004**, 322, 1310-7	3.4	210
671	Autophagy in Huntington disease and huntingtin in autophagy. *Trends in Neurosciences*, **2015**, 38, 26-35	13.3	209
670	Increased ABCA1 activity protects against atherosclerosis. *Journal of Clinical Investigation*, **2002**, 110, 35-42	15.9	203
669	Potent and selective antisense oligonucleotides targeting single-nucleotide polymorphisms in the Huntington disease gene / allele-specific silencing of mutant huntingtin. *Molecular Therapy*, **2011**, 19, 2178-85	11.7	201
668	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, 508-15		199
667	Targeted inactivation of hepatic Abca1 causes profound hypoalphalipoproteinemia and kidney hypercatabolism of apoA-I. *Journal of Clinical Investigation*, **2005**, 115, 1333-42	15.9	190
666	When good drugs go bad. *Nature*, **2007**, 446, 975-7	50.4	188
665	BDNF overexpression in the forebrain rescues Huntington's disease phenotypes in YAC128 mice. *Journal of Neuroscience*, **2010**, 30, 14708-18	6.6	187
664	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, 1503-18	7.3	184
663	Wild-type huntingtin reduces the cellular toxicity of mutant huntingtin in vivo. *American Journal of Human Genetics*, **2001**, 68, 313-24	11	183
662	Clinical markers of early disease in persons near onset of Huntington's disease. *Neurology*, **2001**, 57, 658-62	6.3	180
661	Intramuscular administration of AAV1-lipoprotein lipase S447X lowers triglycerides in lipoprotein lipase-deficient patients. *Arteriosclerosis, Thrombosis, and Vascular Biology*, **2008**, 28, 2303-4	9.4	177
660	Specific caspase interactions and amplification are involved in selective neuronal vulnerability in Huntington's disease. *Cell Death and Differentiation*, **2004**, 11, 424-38	12.7	177
659	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, 43243-56	5.4	176
658	Kennedy's disease: caspase cleavage of the androgen receptor is a crucial event in cytotoxicity. *Journal of Neurochemistry*, **1999**, 72, 185-95	6	173

657	Association between increased arterial-wall thickness and impairment in ABCA1-driven cholesterol efflux: an observational study. *Lancet, The*, **2002**, 359, 37-42	40	171
656	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, 1535-40	5.6	170
655	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, 13318-23	11.5	168
654	CAG expansion in the Huntington disease gene is associated with a specific and targetable predisposing haplogroup. *American Journal of Human Genetics*, **2009**, 84, 351-66	11	163
653	Mutant huntingtin enhances excitotoxic cell death. *Molecular and Cellular Neurosciences*, **2001**, 17, 41-53	4.8	158
652	Beyond disgust: impaired recognition of negative emotions prior to diagnosis in Huntington's disease. *Brain*, **2007**, 130, 1732-44	11.2	156
651	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, 33969-79	5.4	156
650	HIP14, a novel ankyrin domain-containing protein, links huntingtin to intracellular trafficking and endocytosis. *Human Molecular Genetics*, **2002**, 11, 2815-28	5.6	156
649	A coding variant in RARG confers susceptibility to anthracycline-induced cardiotoxicity in childhood cancer. *Nature Genetics*, **2015**, 47, 1079-84	36.3	155
648	Apolipoprotein B gene variants are involved in the determination of serum cholesterol levels: a study in normo- and hyperlipidaemic individuals. *Atherosclerosis*, **1987**, 67, 81-9	3.1	155
647	A highly polymorphic locus very tightly linked to the Huntington's disease gene. *Nature*, **1988**, 332, 734-6	50.4	155
646	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, 2103-14	5.6	154
645	Homozygosity for CAG mutation in Huntington disease is associated with a more severe clinical course. *Brain*, **2003**, 126, 946-55	11.2	151
644	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		151
643	Subtype-specific enhancement of NMDA receptor currents by mutant huntingtin. *Journal of Neurochemistry*, **1999**, 72, 1890-8	6	150
642	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, 929-34	9.4	147
641	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, 469-79	4.8	143
640	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, 65-7	5.6	143

639	Differential effect of the rs4149056 variant in SLCO1B1 on myopathy associated with simvastatin and atorvastatin. *Pharmacogenomics Journal*, **2012**, 12, 233-7	3.5	142
638	Huntingtin bodies sequester vesicle-associated proteins by a polyproline-dependent interaction. *Journal of Neuroscience*, **2004**, 24, 269-81	6.6	142
637	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, 728-34	15.9	142
636	Positron emission tomography in the early diagnosis of Huntington's disease. *Neurology*, **1986**, 36, 888-94	8.5	142
635	HIP1 functions in clathrin-mediated endocytosis through binding to clathrin and adaptor protein 2. *Journal of Biological Chemistry*, **2001**, 276, 39271-6	5.4	141
634	Age-dependent alterations of corticostriatal activity in the YAC128 mouse model of Huntington disease. *Journal of Neuroscience*, **2009**, 29, 2414-27	6.6	139
633	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, 290-7		138
632	Selective degeneration and nuclear localization of mutant huntingtin in the YAC128 mouse model of Huntington disease. *Human Molecular Genetics*, **2005**, 14, 3823-35	5.6	137
631	Automated deformation analysis in the YAC128 Huntington disease mouse model. *NeuroImage*, **2008**, 39, 32-9	7.9	136
630	A genome scan for modifiers of age at onset in Huntington disease: The HD MAPS study. *American Journal of Human Genetics*, **2003**, 73, 682-7	11	131
629	Recommendations for genetic testing to reduce the incidence of anthracycline-induced cardiotoxicity. *British Journal of Clinical Pharmacology*, **2016**, 82, 683-95	3.8	129
628	Small changes, big impact: posttranslational modifications and function of huntingtin in Huntington disease. *Neuroscientist*, **2011**, 17, 475-92	7.6	127
627	Variations on a gene: rare and common variants in ABCA1 and their impact on HDL cholesterol levels and atherosclerosis. *Annual Review of Nutrition*, **2006**, 26, 105-29	9.9	127
626	Loss of wild-type huntingtin influences motor dysfunction and survival in the YAC128 mouse model of Huntington disease. *Human Molecular Genetics*, **2005**, 14, 1379-92	5.6	127
625	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, 217-24		127
624	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, 2738-51	5.6	126
623	Elevated brain 3-hydroxykynurenine and quinolinate levels in Huntington disease mice. *Neurobiology of Disease*, **2006**, 23, 190-7	7.5	125
622	Wild-type huntingtin protects neurons from excitotoxicity. *Journal of Neurochemistry*, **2006**, 96, 1121-9	6	125

621	Predictive, pre-natal and diagnostic genetic testing for Huntington's disease: the experience in Canada from 1987 to 2000. *Clinical Genetics*, **2003**, 63, 462-75	4	125
620	Ethyl-EPA in Huntington disease: a double-blind, randomized, placebo-controlled trial. *Neurology*, **2005**, 65, 286-92	6.5	125
619	Lipoprotein lipase S447X: a naturally occurring gain-of-function mutation. *Arteriosclerosis, Thrombosis, and Vascular Biology*, **2006**, 26, 1236-45	9.4	121
618	Prevention of depressive behaviour in the YAC128 mouse model of Huntington disease by mutation at residue 586 of huntingtin. *Brain*, **2009**, 132, 919-32	11.2	120
617	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, 9634-50	20.1	119
616	Neuronal palmitoyl acyl transferases exhibit distinct substrate specificity. *FASEB Journal*, **2009**, 23, 2605-15	6.9	119
615	Validation of variants in SLC28A3 and UGT1A6 as genetic markers predictive of anthracycline-induced cardiotoxicity in children. *Pediatric Blood and Cancer*, **2013**, 60, 1375-81	3	117
614	Interaction of normal and expanded CAG repeat sizes influences age at onset of Huntington disease **2003**, 119A, 279-82		117
613	Huntingtin associates with acidic phospholipids at the plasma membrane. *Journal of Biological Chemistry*, **2005**, 280, 36464-73	5.4	117
612	Phenotypic variation in heterozygous familial hypercholesterolemia: a comparison of Chinese patients with the same or similar mutations in the LDL receptor gene in China or Canada. *Arteriosclerosis, Thrombosis, and Vascular Biology*, **1998**, 18, 309-15	9.4	117
611	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, 1911-8	5.6	117
610	Rethinking genotype and phenotype correlations in polyglutamine expansion disorders. *Human Molecular Genetics*, **1997**, 6, 2005-10	5.6	116
609	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, 9595-600	11.5	112
608	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, 1064-73	11	111
607	HTT haplotypes contribute to differences in Huntington disease prevalence between Europe and East Asia. *European Journal of Human Genetics*, **2011**, 19, 561-6	5.3	110
606	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, 2219-32	5.6	110
605	Verbal episodic memory declines prior to diagnosis in Huntington's disease. *Neuropsychologia*, **2007**, 45, 1767-76	3.2	109
604	Differential susceptibility to excitotoxic stress in YAC128 mouse models of Huntington disease between initiation and progression of disease. *Journal of Neuroscience*, **2009**, 29, 2193-204	6.6	108

603	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, 1939-51	5.6	108
602	Accurate prediction of the functional significance of single nucleotide polymorphisms and mutations in the ABCA1 gene. *PLoS Genetics*, **2005**, 1, e83	6	107
601	Mitochondrial-dependent Ca2+ handling in Huntington's disease striatal cells: effect of histone deacetylase inhibitors. *Journal of Neuroscience*, **2006**, 26, 11174-86	6.6	106
600	Caspases and neurodegeneration: on the cutting edge of new therapeutic approaches. *Clinical Genetics*, **2000**, 57, 1-10	4	106
599	Deletion of Huntington's disease-linked G8 (D4S10) locus in Wolf-Hirschhorn syndrome. *Nature*, **1985**, 318, 75-8	50.4	106
598	Cholesterol defect is marked across multiple rodent models of Huntington's disease and is manifest in astrocytes. *Journal of Neuroscience*, **2010**, 30, 10844-50	6.6	105
597	Specific loss of brain ABCA1 increases brain cholesterol uptake and influences neuronal structure and function. *Journal of Neuroscience*, **2009**, 29, 3579-89	6.6	105
596	Cholesterol in islet dysfunction and type 2 diabetes. *Journal of Clinical Investigation*, **2008**, 118, 403-8	15.9	105
595	miR-33a modulates ABCA1 expression, cholesterol accumulation, and insulin secretion in pancreatic islets. *Diabetes*, **2012**, 61, 653-8	0.9	104
594	Mitochondrial sensitivity and altered calcium handling underlie enhanced NMDA-induced apoptosis in YAC128 model of Huntington's disease. *Journal of Neuroscience*, **2007**, 27, 13614-23	6.6	104
593	A major insertion accounts for a significant proportion of mutations underlying human lipoprotein lipase deficiency. *Proceedings of the National Academy of Sciences of the United States of America*, **1989**, 86, 948-52	11.5	104
592	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, 1429-35	15.1	103
591	Huntingtin phosphorylation on serine 421 is significantly reduced in the striatum and by polyglutamine expansion in vivo. *Human Molecular Genetics*, **2005**, 14, 1569-77	5.6	103
590	Protein kinase A site-specific phosphorylation regulates ATP-binding cassette A1 (ABCA1)-mediated phospholipid efflux. *Journal of Biological Chemistry*, **2002**, 277, 41835-42	5.4	103
589	Caspase-6 and neurodegeneration. *Trends in Neurosciences*, **2011**, 34, 646-56	13.3	102
588	Activated caspase-6 and caspase-6-cleaved fragments of huntingtin specifically colocalize in the nucleus. *Human Molecular Genetics*, **2008**, 17, 2390-404	5.6	102
587	Full length mutant huntingtin is required for altered Ca2+ signaling and apoptosis of striatal neurons in the YAC mouse model of Huntington's disease. *Neurobiology of Disease*, **2008**, 31, 80-8	7.5	101
586	Multisource ascertainment of Huntington disease in Canada: prevalence and population at risk. *Movement Disorders*, **2014**, 29, 105-14	7	100

585	Enhanced striatal NR2B-containing N-methyl-D-aspartate receptor-mediated synaptic currents in a mouse model of Huntington disease. *Journal of Neurophysiology*, **2004**, 92, 2738-46	3.2	100
584	Absence of disease phenotype and intergenerational stability of the CAG repeat in transgenic mice expressing the human Huntington disease transcript. *Human Molecular Genetics*, **1996**, 5, 177-85	5.6	100
583	Mutant huntingtin N-terminal fragments of specific size mediate aggregation and toxicity in neuronal cells. *Journal of Biological Chemistry*, **2009**, 284, 10855-67	5.4	99
582	Striatal neuronal apoptosis is preferentially enhanced by NMDA receptor activation in YAC transgenic mouse model of Huntington disease. *Neurobiology of Disease*, **2006**, 21, 392-403	7.5	99
581	Cholesterol efflux regulatory protein, Tangier disease and familial high-density lipoprotein deficiency. *Current Opinion in Lipidology*, **2000**, 11, 117-22	4.4	99
580	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, 2877-85	5.6	98
579	Carriers of loss-of-function mutations in ABCA1 display pancreatic beta-cell dysfunction. *Diabetes Care*, **2010**, 33, 869-74	14.6	98
578	HDL and LDL cholesterol significantly influence beta-cell function in type 2 diabetes mellitus. *Current Opinion in Lipidology*, **2010**, 21, 178-85	4.4	98
577	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, 301-9	5.6	98
576	Cortical thickness measured from MRI in the YAC128 mouse model of Huntington's disease. *NeuroImage*, **2008**, 41, 243-51	7.9	98
575	Tissue-specific induction of intestinal ABCA1 expression with a liver X receptor agonist raises plasma HDL cholesterol levels. *Circulation Research*, **2006**, 99, 672-4	15.7	98
574	Psychological consequences and predictors of adverse events in the first 5 years after predictive testing for Huntington's disease. *Clinical Genetics*, **2003**, 64, 300-9	4	97
573	In vivo evaluation of candidate allele-specific mutant huntingtin gene silencing antisense oligonucleotides. *Molecular Therapy*, **2014**, 22, 2093-2106	11.7	96
572	Antisense oligonucleotide therapeutics for inherited neurodegenerative diseases. *Trends in Molecular Medicine*, **2012**, 18, 634-43	11.5	96
571	Elevated plasma triglyceride levels precede amyloid deposition in Alzheimer's disease mouse models with abundant A beta in plasma. *Neurobiology of Disease*, **2006**, 24, 114-27	7.5	96
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