Michael R Hayden

List of Publications by Year in Descending Order

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	Cerebrospinal fluid mutant huntingtin is a biomarker for huntingtin lowering in the striatum of Huntington disease mice.. *Neurobiology of Disease*, **2022**, 105652	7.5	2
745	Neuroprotection of retinal ganglion cells by the sigma-1 receptor agonist pridopidine in models of experimental glaucoma. *Scientific Reports*, **2021**, 11, 21975	4.9	0
744	Sigma-1 and dopamine D2/D3 receptor occupancy of pridopidine in healthy volunteers and patients with Huntington disease: a [F] fluspidine and [F] fallypride PET study. *European Journal of Nuclear Medicine and Molecular Imaging*, **2021**, 48, 1103-1115	8.8	13
743	Sigma-1 Receptor (S1R) Interaction with Cholesterol: Mechanisms of S1R Activation and Its Role in Neurodegenerative Diseases. *International Journal of Molecular Sciences*, **2021**, 22,	6.3	8
742	The Sigma-1 Receptor Mediates Pridopidine Rescue of Mitochondrial Function in Huntington Disease Models. *Neurotherapeutics*, **2021**, 18, 1017-1038	6.4	8
741	Pridopidine reduces mutant huntingtin-induced endoplasmic reticulum stress by modulation of the Sigma-1 receptor. *Journal of Neurochemistry*, **2021**, 158, 467-481	6	6
740	Super-resolution imaging reveals extrastriatal synaptic dysfunction in presymptomatic Huntington disease mice. *Neurobiology of Disease*, **2021**, 152, 105293	7.5	3
739	Reliable Resolution of Full-Length Huntingtin Alleles by Quantitative Immunoblotting. *Journal of Huntingtonjs Disease*, **2021**, 10, 355-365	1.9	2
738	Mutant Huntingtin Is Cleared from the Brain via Active Mechanisms in Huntington Disease. *Journal of Neuroscience*, **2021**, 41, 780-796	6.6	8
737	Rescue of aberrant huntingtin palmitoylation ameliorates mutant huntingtin-induced toxicity. *Neurobiology of Disease*, **2021**, 158, 105479	7.5	3
736	DAPK1 Promotes Extrasynaptic GluN2B Phosphorylation and Striatal Spine Instability in the YAC128 Mouse Model of Huntington Disease. *Frontiers in Cellular Neuroscience*, **2020**, 14, 590569	6.1	5
735	Inhibiting cellular uptake of mutant huntingtin using a monoclonal antibody: Implications for the treatment of Huntington's disease. *Neurobiology of Disease*, **2020**, 141, 104943	7.5	5
734	Compromised IGF signaling causes caspase-6 activation in Huntington disease. *Experimental Neurology*, **2020**, 332, 113396	5.7	3
733	pS421 huntingtin modulates mitochondrial phenotypes and confers neuroprotection in an HD hiPSC model. *Cell Death and Disease*, **2020**, 11, 809	9.8	2
732	Interrupting sequence variants and age of onset in Huntington's disease: clinical implications and emerging therapies. *Lancet Neurology, The*, **2020**, 19, 930-939	24.1	21
731	The Interaction of Aging and Cellular Stress Contributes to Pathogenesis in Mouse and Human Huntington Disease Neurons. *Frontiers in Aging Neuroscience*, **2020**, 12, 524369	5.3	3
730	Tracing the mutated HTT and haplotype of the African ancestor who spread Huntington disease into the Middle East. *Genetics in Medicine*, **2020**, 22, 1903-1908	8.1	1

729	Coupled Control of Distal Axon Integrity and Somal Responses to Axonal Damage by the Palmitoyl Acyltransferase ZDHHC17. *Cell Reports*, **2020**, 33, 108365	10.6	4
728	Frequency of the loss of CAA interruption in the HTT CAG tract and implications for Huntington disease in the reduced penetrance range. *Genetics in Medicine*, **2020**, 22, 2108-2113	8.1	11
727	Gene expression profiles complement the analysis of genomic modifiers of the clinical onset of Huntington disease. *Human Molecular Genetics*, **2020**, 29, 2788-2802	5.6	8
726	Potent and sustained huntingtin lowering via AAV5 encoding miRNA preserves striatal volume and cognitive function in a humanized mouse model of Huntington disease. *Nucleic Acids Research*, **2020**, 48, 36-54	20.1	25
725	Impairment and Restoration of Homeostatic Plasticity in Cultured Cortical Neurons From a Mouse Model of Huntington Disease. *Frontiers in Cellular Neuroscience*, **2019**, 13, 209	6.1	26
724	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	11	67
723	Pridopidine protects neurons from mutant-huntingtin toxicity via the sigma-1 receptor. *Neurobiology of Disease*, **2019**, 129, 118-129	7.5	25
722	Intrinsic mutant HTT-mediated defects in oligodendroglia cause myelination deficits and behavioral abnormalities in Huntington disease. *Proceedings of the National Academy of Sciences of the United States of America*, **2019**, 116, 9622-9627	11.5	26
721	Altered Regulation of Striatal Neuronal -Methyl-D-Aspartate Receptor Trafficking by Palmitoylation in Huntington Disease Mouse Model. *Frontiers in Synaptic Neuroscience*, **2019**, 11, 3	3.5	14
720	Pharmacogenomics of Vincristine-Induced Peripheral Neuropathy Implicates Pharmacokinetic and Inherited Neuropathy Genes. *Clinical Pharmacology and Therapeutics*, **2019**, 105, 402-410	6.1	36
719	In search of a genetic explanation for LDLc variability in an FH family: common SNPs and a rare mutation in explain only part of LDL variability in an FH family. *Journal of Lipid Research*, **2019**, 60, 1733-1740	6.3	1
718	Activation of Caspase-6 Is Promoted by a Mutant Huntingtin Fragment and Blocked by an Allosteric Inhibitor Compound. *Cell Chemical Biology*, **2019**, 26, 1295-1305.e6	8.2	3
717	Pridopidine Induces Functional Neurorestoration Via the Sigma-1 Receptor in a Mouse Model of Parkinson's Disease. *Neurotherapeutics*, **2019**, 16, 465-479	6.4	34
716	A Comprehensive Haplotype-Targeting Strategy for Allele-Specific HTT Suppression in Huntington Disease. *American Journal of Human Genetics*, **2019**, 105, 1112-1125	11	15
715	Pridopidine, a clinic-ready compound, reduces 3,4-dihydroxyphenylalanine-induced dyskinesia in Parkinsonian macaques. *Movement Disorders*, **2019**, 34, 708-716	7	20
714	Safety and efficacy of pridopidine in patients with Huntington's disease (PRIDE-HD): a phase 2, randomised, placebo-controlled, multicentre, dose-ranging study. *Lancet Neurology, The*, **2019**, 18, 165-176	24.1	50
713	Pridopidine stabilizes mushroom spines in mouse models of Alzheimer's disease by acting on the sigma-1 receptor. *Neurobiology of Disease*, **2019**, 124, 489-504	7.5	33
712	Identification of a novel caspase cleavage site in huntingtin that regulates mutant huntingtin clearance. *FASEB Journal*, **2019**, 33, 3190-3197	0.9	12

711	Laquinimod Treatment Improves Myelination Deficits at the Transcriptional and Ultrastructural Levels in the YAC128 Mouse Model of Huntington Disease. *Molecular Neurobiology*, **2019**, 56, 4464-4478	6.2	19
710	Quantification of Motor Function in Huntington Disease Patients Using Wearable Sensor Devices. *Digital Biomarkers*, **2019**, 3, 103-115	7.1	13
709	The molecular epidemiology of Huntington disease is related to intermediate allele frequency and haplotype in the general population. *American Journal of Medical Genetics Part B: Neuropsychiatric Genetics*, **2018**, 177, 346-357	3.5	34
708	Constitutive ablation of caspase-6 reduces the inflammatory response and behavioural changes caused by peripheral pro-inflammatory stimuli. *Cell Death Discovery*, **2018**, 4, 40	6.9	5
707	A whole brain longitudinal study in the YAC128 mouse model of Huntington's disease shows distinct trajectories of neurochemical, structural connectivity and volumetric changes. *Human Molecular Genetics*, **2018**, 27, 2125-2137	5.6	12
706	HACE1 is essential for astrocyte mitochondrial function and influences Huntington disease phenotypes in vivo. *Human Molecular Genetics*, **2018**, 27, 239-253	5.6	12
705	Further Investigation of the Role of and Pharmacogenomic Variants in the Development of Cisplatin-Induced Ototoxicity in Testicular Cancer Patients. *Clinical Cancer Research*, **2018**, 24, 1866-1871	12.9	20
704	Preventing mutant huntingtin proteolysis and intermittent fasting promote autophagy in models of Huntington disease. *Acta Neuropathologica Communications*, **2018**, 6, 16	7.3	30
703	The global spectrum of protein-coding pharmacogenomic diversity. *Pharmacogenomics Journal*, **2018**, 18, 187-195	3.5	46
702	Genetic ablation of Cyp8b1 preserves host metabolic function by repressing steatohepatitis and altering gut microbiota composition. *American Journal of Physiology - Endocrinology and Metabolism* , **2018**, 314, E418-E432	6	13
701	Characterization of subventricular zone-derived progenitor cells from mild and late symptomatic YAC128 mouse model of Huntington's disease. *Biochimica Et Biophysica Acta - Molecular Basis of Disease*, **2018**, 1864, 34-44	6.9	0
700	Therapeutic modulation of the bile acid pool by Cyp8b1 knockdown protects against nonalcoholic fatty liver disease in mice. *FASEB Journal*, **2018**, 32, 3792-3802	0.9	24
699	Altering cortical input unmasks synaptic phenotypes in the YAC128 cortico-striatal co-culture model of Huntington disease. *BMC Biology*, **2018**, 16, 58	7.3	12
698	Huntingtin suppression restores cognitive function in a mouse model of Huntington's disease. *Science Translational Medicine*, **2018**, 10,	17.5	66
697	Large-scale transcriptomic analysis reveals that pridopidine reverses aberrant gene expression and activates neuroprotective pathways in the YAC128 HD mouse. *Molecular Neurodegeneration*, **2018**, 13, 25	19	16
696	Therapeutic approaches to Huntington disease: from the bench to the clinic. *Nature Reviews Drug Discovery*, **2018**, 17, 729-750	64.1	74
695	A human huntingtin SNP alters post-translational modification and pathogenic proteolysis of the protein causing Huntington disease. *Scientific Reports*, **2018**, 8, 8096	4.9	23
694	Pharmacogenomics strategies to optimize treatments for multiple sclerosis: Insights from clinical research. *Progress in Neurobiology*, **2017**, 152, 114-130	10.9	19

693	Author response: Huntington disease reduced penetrance alleles occur at high frequency in the general population. *Neurology*, **2017**, 88, 334-335	6.5	
692	Histone Deacetylase Inhibitors Protect Against Pyruvate Dehydrogenase Dysfunction in Huntington's Disease. *Journal of Neuroscience*, **2017**, 37, 2776-2794	6.6	38
691	Neurodegeneration: Role of repeats in protein clearance. *Nature*, **2017**, 545, 33-34	50.4	3
690	Association Between SLC16A5 Genetic Variation and Cisplatin-Induced Ototoxic Effects in Adult Patients With Testicular Cancer. *JAMA Oncology*, **2017**, 3, 1558-1562	13.4	29
689	Palmitoylation of caspase-6 by HIP14 regulates its activation. *Cell Death and Differentiation*, **2017**, 24, 433-444	12.7	23
688	Pharmacogenomic screening for anthracycline-induced cardiotoxicity in childhood cancer. *British Journal of Clinical Pharmacology*, **2017**, 83, 1143-1145	3.8	9
687	The targetable A1 Huntington disease haplotype has distinct Amerindian and European origins in Latin America. *European Journal of Human Genetics*, **2017**, 25, 332-340	5.3	13
686	ABCA8 Regulates Cholesterol Efflux and High-Density Lipoprotein Cholesterol Levels. *Arteriosclerosis, Thrombosis, and Vascular Biology*, **2017**, 37, 2147-2155	9.4	34
685	A novel humanized mouse model of Huntington disease for preclinical development of therapeutics targeting mutant huntingtin alleles. *Human Molecular Genetics*, **2017**, 26, 1115-1132	5.6	12
684	A pharmacogenetic signature of high response to Copaxone in late-phase clinical-trial cohorts of multiple sclerosis. *Genome Medicine*, **2017**, 9, 50	14.4	14
683	eEF2K inhibition blocks A□2 neurotoxicity by promoting an NRF2 antioxidant response. *Acta Neuropathologica*, **2017**, 133, 101-119	14.3	30
682	The sigma-1 receptor mediates the beneficial effects of pridopidine in a mouse model of Huntington disease. *Neurobiology of Disease*, **2017**, 97, 46-59	7.5	79
681	Comparative Mitochondrial-Based Protective Effects of Resveratrol and Nicotinamide in Huntington's Disease Models. *Molecular Neurobiology*, **2017**, 54, 5385-5399	6.2	77
680	Epidemiology of Huntington disease. *Handbook of Clinical Neurology / Edited By P J Vinken and G W Bruyn*, **2017**, 144, 31-46	3	30
679	Compositional differences between Copaxone and Glatopa are reflected in altered immunomodulation ex vivo in a mouse model. *Annals of the New York Academy of Sciences*, **2017**, 1407, 75-89	6.5	5
678	Early pridopidine treatment improves behavioral and transcriptional deficits in YAC128 Huntington disease mice. *JCI Insight*, **2017**, 2,	9.9	30
677	An enhanced Q175 knock-in mouse model of Huntington disease with higher mutant huntingtin levels and accelerated disease phenotypes. *Human Molecular Genetics*, **2016**, 25, 3654-3675	5.6	39
676	Huntington disease reduced penetrance alleles occur at high frequency in the general population. *Neurology*, **2016**, 87, 282-8	6.5	48

675	Laquinimod rescues striatal, cortical and white matter pathology and results in modest behavioural improvements in the YAC128 model of Huntington disease. *Scientific Reports*, **2016**, 6, 31652	4.9	43
674	Modeling Doxorubicin-Induced Cardiotoxicity in Human Pluripotent Stem Cell Derived-Cardiomyocytes. *Scientific Reports*, **2016**, 6, 25333	4.9	90
673	Sudden death due to paralysis and synaptic and behavioral deficits when Hip14/Zdhhc17 is deleted in adult mice. *BMC Biology*, **2016**, 14, 108	7.3	13
672	Structural and molecular myelination deficits occur prior to neuronal loss in the YAC128 and BACHD models of Huntington disease. *Human Molecular Genetics*, **2016**, 25, 2621-2632	5.6	40
671	ABCA1 deficiency and cellular cholesterol accumulation increases islet amyloidogenesis in mice. *Diabetologia*, **2016**, 59, 1242-6	10.3	18
670	Treatment with the MAO-A inhibitor clorgyline elevates monoamine neurotransmitter levels and improves affective phenotypes in a mouse model of Huntington disease. *Experimental Neurology*, **2016**, 278, 4-10	5.7	32
669	Design, Characterization, and Lead Selection of Therapeutic miRNAs Targeting Huntingtin for Development of Gene Therapy for Huntington's Disease. *Molecular Therapy - Nucleic Acids*, **2016**, 5, e297	10.7	60
668	Interactome network analysis identifies multiple caspase-6 interactors involved in the pathogenesis of HD. *Human Molecular Genetics*, **2016**, 25, 1600-18	5.6	12
667	Insulin and IGF-1 regularize energy metabolites in neural cells expressing full-length mutant huntingtin. *Neuropeptides*, **2016**, 58, 73-81	3.3	28
666	A true mentor and pioneer in medical genetics. *South African Medical Journal*, **2016**, 106, S7-9	1.5	
665	Genetic diversity of variants involved in drug response and metabolism in Sri Lankan populations: implications for clinical implementation of pharmacogenomics. *Pharmacogenetics and Genomics*, **2016**, 26, 28-39	1.9	14
664	A novel microdeletion affecting the CETP gene raises HDL-associated cholesterol levels. *Clinical Genetics*, **2016**, 89, 495-500	4	
663	Laquinimod decreases Bax expression and reduces caspase-6 activation in neurons. *Experimental Neurology*, **2016**, 283, 121-8	5.7	20
662	B42 Early olfactory behaviour deficits associated with olfactory bulb atrophy and caspase-8 activation in HD rodent models. *Journal of Neurology, Neurosurgery and Psychiatry*, **2016**, 87, A24.1-A24	5.5	
661	Dopamine D2 receptor gene variants and response to rasagiline in early Parkinson's disease: a pharmacogenetic study. *Brain*, **2016**, 139, 2050-62	11.2	38
660	Functional effects of the antigen glatiramer acetate are complex and tightly associated with its composition. *Journal of Neuroimmunology*, **2016**, 290, 84-95	3.5	14
659	Enhanced immune response to MMP3 stimulation in microglia expressing mutant huntingtin. *Neuroscience*, **2016**, 325, 74-88	3.9	22
658	Laquinimod arrests experimental autoimmune encephalomyelitis by activating the aryl hydrocarbon receptor. *Proceedings of the National Academy of Sciences of the United States of America*, **2016**, 113, E6145-E6152	11.5	74

657	Pridopidine activates neuroprotective pathways impaired in Huntington Disease. *Human Molecular Genetics*, **2016**, 25, 3975-3987	5.6	50
656	Recommendations for genetic testing to reduce the incidence of anthracycline-induced cardiotoxicity. *British Journal of Clinical Pharmacology*, **2016**, 82, 683-95	3.8	129
655	Laquinimod dampens hyperactive cytokine production in Huntington's disease patient myeloid cells. *Journal of Neurochemistry*, **2016**, 137, 782-94	6	26
654	A Huntingtin-based peptide inhibitor of caspase-6 provides protection from mutant Huntingtin-induced motor and behavioral deficits. *Human Molecular Genetics*, **2015**, 24, 2604-14	5.6	38
653	Genetic variants in SLC22A17 and SLC22A7 are associated with anthracycline-induced cardiotoxicity in children. *Pharmacogenomics*, **2015**, 16, 1065-76	2.6	73
652	A coding variant in RARG confers susceptibility to anthracycline-induced cardiotoxicity in childhood cancer. *Nature Genetics*, **2015**, 47, 1079-84	36.3	155
651	Systematic interaction network filtering identifies CRMP1 as a novel suppressor of huntingtin misfolding and neurotoxicity. *Genome Research*, **2015**, 25, 701-13	9.7	17
650	A SNP in the HTT promoter alters NF-B binding and is a bidirectional genetic modifier of Huntington disease. *Nature Neuroscience*, **2015**, 18, 807-16	25.5	70
649	Huntington disease. *Nature Reviews Disease Primers*, **2015**, 1, 15005	51.1	672
648	HD iPSC-derived neural progenitors accumulate in culture and are susceptible to BDNF withdrawal due to glutamate toxicity. *Human Molecular Genetics*, **2015**, 24, 3257-71	5.6	74
647	Post-translational myristoylation at the cross roads of cell death, autophagy and neurodegeneration. *Biochemical Society Transactions*, **2015**, 43, 229-34	5.1	17
646	Huntingtin Haplotypes Provide Prioritized Target Panels for Allele-specific Silencing in Huntington Disease Patients of European Ancestry. *Molecular Therapy*, **2015**, 23, 1759-1771	11.7	54
645	Ultrasensitive measurement of huntingtin protein in cerebrospinal fluid demonstrates increase with Huntington disease stage and decrease following brain huntingtin suppression. *Scientific Reports*, **2015**, 5, 12166	4.9	60
644	Targeted next-generation sequencing to diagnose disorders of HDL cholesterol. *Journal of Lipid Research*, **2015**, 56, 1993-2001	6.3	24
643	Comment on Rickels et al. Loss-of-Function Mutations in ABCA1 and Enhanced Cell Secretory Capacity in Young Adults. Diabetes 2015;64:193-199. *Diabetes*, **2015**, 64, e25-6; discussion e27	0.9	2
642	A new mutation for Huntington disease following maternal transmission of an intermediate allele. *European Journal of Medical Genetics*, **2015**, 58, 28-30	2.6	15
641	Inhibition of Excessive Monoamine Oxidase A/B Activity Protects Against Stress-induced Neuronal Death in Huntington Disease. *Molecular Neurobiology*, **2015**, 52, 1850-1861	6.2	25
640	Autophagy in Huntington disease and huntingtin in autophagy. *Trends in Neurosciences*, **2015**, 38, 26-35	13.3	209

639	Aberrant palmitoylation in Huntington disease. *Biochemical Society Transactions*, **2015**, 43, 205-10	5.1	21
638	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	5	69
637	Gene expression studies of a human monocyte cell line identify dissimilarities between differently manufactured glatiramoids. *Scientific Reports*, **2015**, 5, 10191	4.9	12
636	A longitudinal study of magnetic resonance spectroscopy Huntington's disease biomarkers. *Movement Disorders*, **2015**, 30, 393-401	7	38
635	Direct intracerebral delivery of a miR-33 antisense oligonucleotide into mouse brain increases brain ABCA1 expression. [Corrected]. *Neuroscience Letters*, **2015**, 598, 66-72	3.3	24
634	Loss of Cyp8b1 improves glucose homeostasis by increasing GLP-1. *Diabetes*, **2015**, 64, 1168-79	0.9	60
633	Huntingtin interacting proteins 14 and 14-like are required for chorioallantoic fusion during early placental development. *Developmental Biology*, **2015**, 397, 257-66	3.1	7
632	Partial rescue of some features of Huntington Disease in the genetic absence of caspase-6 in YAC128 mice. *Neurobiology of Disease*, **2015**, 76, 24-36	7.5	39
631	Anti-semaphorin 4D immunotherapy ameliorates neuropathology and some cognitive impairment in the YAC128 mouse model of Huntington disease. *Neurobiology of Disease*, **2015**, 76, 46-56	7.5	60
630	Biophysical and biological characterization of hairpin and molecular beacon RNase H active antisense oligonucleotides. *ACS Chemical Biology*, **2015**, 10, 1227-33	4.9	9
629	Human genetics of HDL: Insight into particle metabolism and function. *Progress in Lipid Research*, **2015**, 58, 14-25	14.3	35
628	Clinical, Biochemical, and Molecular Characterization of Novel Mutations in ABCA1 in Families with Tangier Disease. *JIMD Reports*, **2015**, 18, 51-62	1.9	15
627	Evidence-based genetic counselling implications for Huntington disease intermediate allele predictive test results. *Clinical Genetics*, **2014**, 85, 303-11	4	27
626	Personalized gene silencing therapeutics for Huntington disease. *Clinical Genetics*, **2014**, 86, 29-36	4	39
625	Codeine-related deaths: The role of pharmacogenetics and drug interactions. *Forensic Science International*, **2014**, 239, 50-6	2.6	33
624	High density lipoprotein metabolism in low density lipoprotein receptor-deficient mice. *Journal of Lipid Research*, **2014**, 55, 1914-24	6.3	14
623	Pharmacogenomic diversity in Singaporean populations and Europeans. *Pharmacogenomics Journal*, **2014**, 14, 555-63	3.5	14
622	Identification of four novel genes contributing to familial elevated plasma HDL cholesterol in humans. *Journal of Lipid Research*, **2014**, 55, 1693-701	6.3	17

621	ABCA1 in adipocytes regulates adipose tissue lipid content, glucose tolerance, and insulin sensitivity. *Journal of Lipid Research*, **2014**, 55, 516-23	6.3	61
620	Use of genetic technologies to compare medicines. *Clinical Genetics*, **2014**, 86, 441-6	4	
619	Public perceptions of pharmacogenetics. *Pediatrics*, **2014**, 133, e1258-67	7.4	19
618	Response to "evaluation of pharmacogenetic markers to predict the risk of Cisplatin-induced ototoxicity". *Clinical Pharmacology and Therapeutics*, **2014**, 96, 158	6.1	2
617	Laquinimod exerts strong clinical and immunomodulatory effects in Lewis rat experimental autoimmune neuritis. *Journal of Neuroimmunology*, **2014**, 274, 38-45	3.5	15
616	Genetic markers of cisplatin-induced hearing loss in children. *Clinical Pharmacology and Therapeutics*, **2014**, 96, 296-8	6.1	7
615	VKORC1 and CYP2C9 genotypes are predictors of warfarin-related outcomes in children. *Pediatric Blood and Cancer*, **2014**, 61, 1055-62	3	31
614	Multisource ascertainment of Huntington disease in Canada: prevalence and population at risk. *Movement Disorders*, **2014**, 29, 105-14	7	100
613	p53 increases caspase-6 expression and activation in muscle tissue expressing mutant huntingtin. *Human Molecular Genetics*, **2014**, 23, 717-29	5.6	38
612	HACE1 reduces oxidative stress and mutant Huntingtin toxicity by promoting the NRF2 response. *Proceedings of the National Academy of Sciences of the United States of America*, **2014**, 111, 3032-7	11.5	65
611	Higher frequency of genetic variants conferring increased risk for ADRs for commonly used drugs treating cancer, AIDS and tuberculosis in persons of African descent. *Pharmacogenomics Journal*, **2014**, 14, 160-70	3.5	23
610	Identification of a post-translationally myristoylated autophagy-inducing domain released by caspase cleavage of huntingtin. *Human Molecular Genetics*, **2014**, 23, 3166-79	5.6	43
609	Hepatic ABCA1 expression improves □cell function and glucose tolerance. *Diabetes*, **2014**, 63, 4076-82	0.9	16
608	Two novel mutations in apolipoprotein C3 underlie atheroprotective lipid profiles in families. *Clinical Genetics*, **2014**, 85, 433-40	4	15
607	In vivo evaluation of candidate allele-specific mutant huntingtin gene silencing antisense oligonucleotides. *Molecular Therapy*, **2014**, 22, 2093-2106	11.7	96
606	Role of TPMT and COMT genetic variation in cisplatin-induced ototoxicity. *Clinical Pharmacology and Therapeutics*, **2014**, 95, 253	6.1	18
605	The emerging era of pharmacogenomics: current successes, future potential, and challenges. *Clinical Genetics*, **2014**, 86, 21-8	4	54
604	Development of a broad-based ADME panel for use in pharmacogenomic studies. *Pharmacogenomics*, **2014**, 15, 1185-95	2.6	8

603	Bidirectional control of postsynaptic density-95 (PSD-95) clustering by Huntingtin. *Journal of Biological Chemistry*, **2014**, 289, 3518-28	5.4	23
602	The palmitoyl acyltransferase HIP14 shares a high proportion of interactors with huntingtin: implications for a role in the pathogenesis of Huntington's disease. *Human Molecular Genetics*, **2014**, 23, 4142-60	5.6	44
601	IGF-1 intranasal administration rescues Huntington's disease phenotypes in YAC128 mice. *Molecular Neurobiology*, **2014**, 49, 1126-42	6.2	50
600	Comparing the biological impact of glatiramer acetate with the biological impact of a generic. *PLoS ONE*, **2014**, 9, e83757	3.7	26
599	Identification of binding sites in Huntingtin for the Huntingtin Interacting Proteins HIP14 and HIP14L. *PLoS ONE*, **2014**, 9, e90669	3.7	18
598	Striatal synaptic dysfunction and hippocampal plasticity deficits in the Hu97/18 mouse model of Huntington disease. *PLoS ONE*, **2014**, 9, e94562	3.7	29
597	Allele-specific suppression of mutant huntingtin using antisense oligonucleotides: providing a therapeutic option for all Huntington disease patients. *PLoS ONE*, **2014**, 9, e107434	3.7	81
596	"Grasping the grey": patient understanding and interpretation of an intermediate allele predictive test result for Huntington disease. *Journal of Genetic Counseling*, **2013**, 22, 200-17	2.5	21
595	Replication of TPMT and ABCC3 genetic variants highly associated with cisplatin-induced hearing loss in children. *Clinical Pharmacology and Therapeutics*, **2013**, 94, 243-51	6.1	87
594	Suppressing aberrant GluN3A expression rescues synaptic and behavioral impairments in Huntington's disease models. *Nature Medicine*, **2013**, 19, 1030-8	50.5	79
593	Hunting human disease genes: lessons from the past, challenges for the future. *Human Genetics*, **2013**, 132, 603-17	6.3	30
592	HLA-A 31:01 and HLA-B 15:02 as genetic markers for carbamazepine hypersensitivity in children. *Clinical Pharmacology and Therapeutics*, **2013**, 94, 142-9	6.1	91
591	Huntington disease in the South African population occurs on diverse and ethnically distinct genetic haplotypes. *European Journal of Human Genetics*, **2013**, 21, 1120-7	5.3	37
590	CAG size-specific risk estimates for intermediate allele repeat instability in Huntington disease. *Journal of Medical Genetics*, **2013**, 50, 696-703	5.8	50
589	A systematic review and meta-analysis of clinical variables used in Huntington disease research. *Movement Disorders*, **2013**, 28, 1987-94	7	6
588	Choosing an animal model for the study of Huntington's disease. *Nature Reviews Neuroscience*, **2013**, 14, 708-21	13.5	227
587	Candidate glutamatergic and dopaminergic pathway gene variants do not influence Huntington's disease motor onset. *Neurogenetics*, **2013**, 14, 173-9	3	9
586	High frequency of intermediate alleles on Huntington disease-associated haplotypes in British Columbia's general population. *American Journal of Medical Genetics Part B: Neuropsychiatric Genetics*, **2013**, 162B, 864-71	3.5	29

585	Hip14l-deficient mice develop neuropathological and behavioural features of Huntington disease. *Human Molecular Genetics*, **2013**, 22, 452-65	5.6	51
584	Tracking brain palmitoylation change: predominance of glial change in a mouse model of Huntington's disease. *Chemistry and Biology*, **2013**, 20, 1421-34		63
583	ABCA1 influences neuroinflammation and neuronal death. *Neurobiology of Disease*, **2013**, 54, 445-55	7.5	43
582	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
581	Cancer pharmacogenomics in children: research initiatives and progress to date. *Paediatric Drugs*, **2013**, 15, 71-81	4.2	5
580	From mutation identification to therapy: discovery and origins of the first approved gene therapy in the Western world. *Human Gene Therapy*, **2013**, 24, 472-8	4.8	35
579	A fully humanized transgenic mouse model of Huntington disease. *Human Molecular Genetics*, **2013**, 22, 18-34	5.6	66
578	Developing a comprehensive, effective patient-friendly website to enhance decision making in predictive testing for Huntington disease. *Genetics in Medicine*, **2013**, 15, 466-72	8.1	6
577	When access is an issue: exploring barriers to predictive testing for Huntington disease in British Columbia, Canada. *European Journal of Human Genetics*, **2013**, 21, 148-53	5.3	18
576	Intrinsic cleavage of receptor-interacting protein kinase-1 by caspase-6. *Cell Death and Differentiation*, **2013**, 20, 86-96	12.7	30
575	The Salmonella type III effector SspH2 specifically exploits the NLR co-chaperone activity of SGT1 to subvert immunity. *PLoS Pathogens*, **2013**, 9, e1003518	7.6	57
574	The impact of partial and complete loss-of-function mutations in endothelial lipase on high-density lipoprotein levels and functionality in humans. *Circulation: Cardiovascular Genetics*, **2013**, 6, 54-62		46
573	Regulation of ABCA1 protein expression and function in hepatic and pancreatic islet cells by miR-145. *Arteriosclerosis, Thrombosis, and Vascular Biology*, **2013**, 33, 2724-32	9.4	85
572	Putative association of ABCB1 2677G>T/A with oxycodone-induced central nervous system depression in breastfeeding mothers. *Therapeutic Drug Monitoring*, **2013**, 35, 466-72	3.2	14
571	De novo Huntington disease caused by 26-44 CAG repeat expansion on a low-risk haplotype. *Neurology*, **2013**, 81, 1099-100	6.5	7
570	Memory and synaptic deficits in Hip14/DHHC17 knockout mice. *Proceedings of the National Academy of Sciences of the United States of America*, **2013**, 110, 20296-301	11.5	41
569	Providing predictive testing for Huntington disease via telehealth: results of a pilot study in British Columbia, Canada. *Clinical Genetics*, **2013**, 84, 60-4	4	23
568	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

567	Antisense oligonucleotide-mediated correction of transcriptional dysregulation is correlated with behavioral benefits in the YAC128 mouse model of Huntington's disease. *Journal of Huntingtonjs Disease*, **2013**, 2, 217-28	1.9	49
566	Life-threatening adverse events following therapeutic opioid administration in adults: is pharmacogenetic analysis useful?. *Pain Research and Management*, **2013**, 18, 133-6	2.6	12
565	A clinical tool for reducing central nervous system depression among neonates exposed to codeine through breast milk. *PLoS ONE*, **2013**, 8, e70073	3.7	27
564	Pharmacogenomics of vincristine-induced neurotoxicity in pediatric cancer patients. *FASEB Journal*, **2013**, 27, 666.3	0.9	
563	An investigation of morphine-to-codeine metabolic ratios in postmortem blood, drug interactions, and cytochrome P450 2D6 (CYP2D6) genotype. *FASEB Journal*, **2013**, 27, 666.2	0.9	
562	Predicting Anthracycline-induced Cardiotoxicity in Children Genome-Wide Association Study. *FASEB Journal*, **2013**, 27, 663.3	0.9	
561	Central nervous system depression of neonates breastfed by mothers receiving oxycodone for postpartum analgesia. *Journal of Pediatrics*, **2012**, 160, 33-7.e2	3.6	81
560	Age-dependent neurovascular abnormalities and altered microglial morphology in the YAC128 mouse model of Huntington disease. *Neurobiology of Disease*, **2012**, 45, 438-49	7.5	79
559	Treatment of Na(v)1.7-mediated pain in inherited erythromelalgia using a novel sodium channel blocker. *Pain*, **2012**, 153, 80-85	8	94
558	Beyond the patient: the broader impact of genetic discrimination among individuals at risk of Huntington disease. *American Journal of Medical Genetics Part B: Neuropsychiatric Genetics*, **2012**, 159B, 217-26	3.5	32
557	Human Mendelian pain disorders: a key to discovery and validation of novel analgesics. *Clinical Genetics*, **2012**, 82, 367-73	4	30
556	CAG repeat expansion in Huntington disease determines age at onset in a fully dominant fashion. *Neurology*, **2012**, 78, 690-5	6.5	231
555	Polyglutamine diseases and the risk of cancer. *Lancet Oncology, The*, **2012**, 13, 569-71	21.7	1
554	Population stratification may bias analysis of PGC-1 as a modifier of age at Huntington disease motor onset. *Human Genetics*, **2012**, 131, 1833-40	6.3	25
553	Putting proteins in their place: palmitoylation in Huntington disease and other neuropsychiatric diseases. *Progress in Neurobiology*, **2012**, 97, 220-38	10.9	95
552	Increased risk of coronary artery disease in Caucasians with extremely low HDL cholesterol due to mutations in ABCA1, APOA1, and LCAT. *Biochimica Et Biophysica Acta - Molecular and Cell Biology of Lipids*, **2012**, 1821, 416-24	5	38
551	TAA repeat variation in the GRIK2 gene does not influence age at onset in Huntington's disease. *Biochemical and Biophysical Research Communications*, **2012**, 424, 404-8	3.4	17
550	More codeine fatalities after tonsillectomy in North American children. *Pediatrics*, **2012**, 129, e1343-7	7.4	289

549	Antisense oligonucleotide therapeutics for inherited neurodegenerative diseases. *Trends in Molecular Medicine*, **2012**, 18, 634-43	11.5	96
548	NP03, a novel low-dose lithium formulation, is neuroprotective in the YAC128 mouse model of Huntington disease. *Neurobiology of Disease*, **2012**, 48, 282-9	7.5	42
547	Apnea and Oxygen Desaturations in Children Treated with Opioids after Adenotonsillectomy for Obstructive Sleep Apnea Syndrome. *Paediatric Drugs*, **2012**, 14, 411-415	4.2	24
546	Differential effect of the rs4149056 variant in SLCO1B1 on myopathy associated with simvastatin and atorvastatin. *Pharmacogenomics Journal*, **2012**, 12, 233-7	3.5	142
545	Age-Dependent Resistance to Excitotoxicity in Htt CAG140 Mice and the Effect of Strain Background. *Journal of Huntingtonjs Disease*, **2012**, 1, 221-41	1.9	7
544	Economic impact of a genetic test for cisplatin-induced ototoxicity. *Pharmacogenomics Journal*, **2012**, 12, 205-13	3.5	16
543	Loss of both ABCA1 and ABCG1 results in increased disturbances in islet sterol homeostasis, inflammation, and impaired Ecell function. *Diabetes*, **2012**, 61, 659-64	0.9	85
542	Medicine. Whole-genome sequencing: the new standard of care?. *Science*, **2012**, 336, 1112-3	33.3	58
541	Common SNP-based haplotype analysis of the 4p16.3 Huntington disease gene region. *American Journal of Human Genetics*, **2012**, 90, 434-44	11	48
540	Adoption and the communication of genetic risk: experiences in Huntington disease. *Clinical Genetics*, **2012**, 81, 64-9	4	5
539	What monozygotic twins discordant for phenotype illustrate about mechanisms influencing genetic forms of neurodegeneration. *Clinical Genetics*, **2012**, 81, 325-33	4	26
538	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
537	Postpartum maternal codeine therapy and the risk of adverse neonatal outcomes: the devil is in the details. *Therapeutic Drug Monitoring*, **2012**, 34, 378-80	3.2	9
536	Rescue from excitotoxicity and axonal degeneration accompanied by age-dependent behavioral and neuroanatomical alterations in caspase-6-deficient mice. *Human Molecular Genetics*, **2012**, 21, 1954-67	5.6	58
535	Peripheral and cerebral metabolic features in an animal model of Huntington's disease **2012**,		1
534	miR-33a modulates ABCA1 expression, cholesterol accumulation, and insulin secretion in pancreatic islets. *Diabetes*, **2012**, 61, 653-8	0.9	104
533	Pharmacogenomic prediction of anthracycline-induced cardiotoxicity in children. *Journal of Clinical Oncology*, **2012**, 30, 1422-8	2.2	267
532	Transgenic mouse model expressing the caspase 6 fragment of mutant huntingtin. *Journal of Neuroscience*, **2012**, 32, 183-93	6.6	44

531	Caspase-6-Resistant Mutant Huntingtin Does not Rescue the Toxic Effects of Caspase-Cleavable Mutant Huntingtin in vivo. *Journal of Huntingtonjs Disease*, **2012**, 1, 243-60	1.9	7
530	Personalized Medicine: Temper Expectations--Response. *Science*, **2012**, 337, 911-911	33.3	2
529	Prediction of codeine toxicity in infants and their mothers using a novel combination of maternal genetic markers. *Clinical Pharmacology and Therapeutics*, **2012**, 91, 692-9	6.1	52
528	Low levels of human HIP14 are sufficient to rescue neuropathological, behavioural, and enzymatic defects due to loss of murine HIP14 in Hip14-/- mice. *PLoS ONE*, **2012**, 7, e36315	3.7	9
527	Segregation of LIPG, CETP, and GALNT2 mutations in Caucasian families with extremely high HDL cholesterol. *PLoS ONE*, **2012**, 7, e37437	3.7	26
526	Apnea and oxygen desaturations in children treated with opioids after adenotonsillectomy for obstructive sleep apnea syndrome: a prospective pilot study. *Paediatric Drugs*, **2012**, 14, 411-5	4.2	7
525	Small changes, big impact: posttranslational modifications and function of huntingtin in Huntington disease. *Neuroscientist*, **2011**, 17, 475-92	7.6	127
524	Convergent pathogenic pathways in Alzheimer's and Huntington's diseases: shared targets for drug development. *Nature Reviews Drug Discovery*, **2011**, 10, 853-67	64.1	58
523	Altered palmitoylation and neuropathological deficits in mice lacking HIP14. *Human Molecular Genetics*, **2011**, 20, 3899-909	5.6	90
522	The dynamics of macrophage infiltration into the arterial wall during atherosclerotic lesion development in low-density lipoprotein receptor knockout mice. *American Journal of Pathology*, **2011**, 178, 413-22	5.8	19
521	Islet cholesterol accumulation due to loss of ABCA1 leads to impaired exocytosis of insulin granules. *Diabetes*, **2011**, 60, 3186-96	0.9	76
520	Caspase-6 and neurodegeneration. *Trends in Neurosciences*, **2011**, 34, 646-56	13.3	102
519	Lessons from predictive testing for Huntington disease: 25 years on. *Journal of Medical Genetics*, **2011**, 48, 649-50	5.8	16
518	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
517	Cholesterol metabolism in Huntington disease. *Nature Reviews Neurology*, **2011**, 7, 561-72	15	94
516	Amelioration of hypertriglyceridemia with hypo-alpha-cholesterolemia in LPL deficient mice by hematopoietic cell-derived LPL. *PLoS ONE*, **2011**, 6, e25620	3.7	4
515	CYP2D6 polymorphisms and codeine analgesia in postpartum pain management: a pilot study. *Therapeutic Drug Monitoring*, **2011**, 33, 425-32	3.2	35
514	Pharmacogenomics of cardiovascular drugs and adverse effects in pediatrics. *Journal of Cardiovascular Pharmacology*, **2011**, 58, 228-39	3.1	16

513	Novel mutations in scavenger receptor BI associated with high HDL cholesterol in humans. *Clinical Genetics*, **2011**, 79, 575-81	4	60
512	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
511	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
510	Altered adult hippocampal neurogenesis in the YAC128 transgenic mouse model of Huntington disease. *Neurobiology of Disease*, **2011**, 41, 249-60	7.5	86
509	Natural history of disease in the YAC128 mouse reveals a discrete signature of pathology in Huntington disease. *Neurobiology of Disease*, **2011**, 43, 257-65	7.5	56
508	Mice lacking caspase-2 are protected from behavioral changes, but not pathology, in the YAC128 model of Huntington disease. *Molecular Neurodegeneration*, **2011**, 6, 59	19	18
507	Factors associated with experiences of genetic discrimination among individuals at risk for Huntington disease. *American Journal of Medical Genetics Part B: Neuropsychiatric Genetics*, **2011**, 156B, 19-27	3.5	14
506	Diagnostic testing for vaccinomics: is the regulatory approval framework adequate? A comparison of Canada, the United States, and Europe. *OMICS A Journal of Integrative Biology*, **2011**, 15, 597-605	3.8	2
505	A grand challenge: providing benefits of clinical genetics to those in need. *Genetics in Medicine*, **2011**, 13, 197-200	8.1	39
504	Wild-type HTT modulates the enzymatic activity of the neuronal palmitoyl transferase HIP14. *Human Molecular Genetics*, **2011**, 20, 3356-65	5.6	61
503	A quantitative method for the specific assessment of caspase-6 activity in cell culture. *PLoS ONE*, **2011**, 6, e27680	3.7	24
502	Communicating pharmacogenetic research results to breastfeeding mothers taking codeine: a pilot study of perceptions and benefits. *Clinical Pharmacology and Therapeutics*, **2010**, 88, 792-5	6.1	21
501	BDNF overexpression in the forebrain rescues Huntington's disease phenotypes in YAC128 mice. *Journal of Neuroscience*, **2010**, 30, 14708-18	6.6	187
500	Transcriptional changes in Huntington disease identified using genome-wide expression profiling and cross-platform analysis. *Human Molecular Genetics*, **2010**, 19, 1438-52	5.6	74
499	Full-length huntingtin levels modulate body weight by influencing insulin-like growth factor 1 expression. *Human Molecular Genetics*, **2010**, 19, 1528-38	5.6	79
498	Glucose intolerance and decreased early insulin response in mice with severe hypertriglyceridemia. *Experimental Biology and Medicine*, **2010**, 235, 40-6	3.7	7
497	Mutation of conserved cysteines in the Ly6 domain of GPIHBP1 in familial chylomicronemia. *Journal of Lipid Research*, **2010**, 51, 1535-45	6.3	90
496	Magnetic resonance spectroscopy biomarkers in premanifest and early Huntington disease. *Neurology*, **2010**, 75, 1702-10	6.5	84

495	Targeted deletion of hepatocyte ABCA1 leads to very low density lipoprotein triglyceride overproduction and low density lipoprotein hypercatabolism. *Journal of Biological Chemistry*, **2010**, 285, 12197-209	5.4	50
494	The Canadian Pharmacogenomics Network for Drug Safety: a model for safety pharmacology. *Thyroid*, **2010**, 20, 681-7	6.2	49
493	Fatal hydrocodone overdose in a child: pharmacogenetics and drug interactions. *Pediatrics*, **2010**, 126, e986-9	7.4	75
492	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
491	Phosphorylation of huntingtin at Ser421 in YAC128 neurons is associated with protection of YAC128 neurons from NMDA-mediated excitotoxicity and is modulated by PP1 and PP2A. *Journal of Neuroscience*, **2010**, 30, 14318-29	6.6	69
490	Cleavage at the 586 amino acid caspase-6 site in mutant huntingtin influences caspase-6 activation in vivo. *Journal of Neuroscience*, **2010**, 30, 15019-29	6.6	85
489	Carriers of loss-of-function mutations in ABCA1 display pancreatic beta-cell dysfunction. *Diabetes Care*, **2010**, 33, 869-74	14.6	98
488	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
487	Pharmacogenomics and active surveillance for serious adverse drug reactions in children. *Pharmacogenomics*, **2010**, 11, 1269-85	2.6	16
486	Cases: Cocaine adulterant linked to neutropenia. *Cmaj*, **2010**, 182, 57-9	3.5	23
485	The metabolic phenotype of SCD1-deficient mice is independent of melanin-concentrating hormone. *Peptides*, **2010**, 31, 123-9	3.8	2
484	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
483	Mycophenolate mofetil attenuates plaque inflammation in patients with symptomatic carotid artery stenosis. *Atherosclerosis*, **2010**, 211, 231-6	3.1	50
482	Unstable familial transmissions of Huntington disease alleles with 27-35 CAG repeats (intermediate alleles). *American Journal of Medical Genetics Part B: Neuropsychiatric Genetics*, **2010**, 153B, 314-20	3.5	19
481	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
480	Cholesterol efflux via ATP-binding cassette transporter A1 (ABCA1) and cholesterol uptake via the LDL receptor influences cholesterol-induced impairment of beta cell function in mice. *Diabetologia*, **2010**, 53, 1110-9	10.3	92
479	Cholesterol toxicity in pancreatic islets from LDL receptor-deficient mice. Reply to: de Souza JC, de Oliveira CAM, Carneiro EM et al. [letter]. *Diabetologia*, **2010**, 53, 2463-2464	10.3	1
478	Cholesterol in beta-cell dysfunction: the emerging connection between HDL cholesterol and type 2 diabetes. *Current Diabetes Reports*, **2010**, 10, 55-60	5.6	46

477	Palmitoylation and function of glial glutamate transporter-1 is reduced in the YAC128 mouse model of Huntington disease. *Neurobiology of Disease*, **2010**, 40, 207-15	7.5	87
476	Adenosine-triphosphate-binding cassette transporter-1 trafficking and function. *Trends in Cardiovascular Medicine*, **2010**, 20, 41-9	6.9	34
475	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
474	Perception, experience, and response to genetic discrimination in Huntington disease: the international RESPOND-HD study. *American Journal of Medical Genetics Part B: Neuropsychiatric Genetics*, **2010**, 153B, 1081-93	3.5	27
473	In their own words: reports of stigma and genetic discrimination by people at risk for Huntington disease in the International RESPOND-HD study. *American Journal of Medical Genetics Part B: Neuropsychiatric Genetics*, **2010**, 153B, 1150-9	3.5	24
472	Interaction of postsynaptic density protein-95 with NMDA receptors influences excitotoxicity in the yeast artificial chromosome mouse model of Huntington's disease. *Journal of Neuroscience*, **2009**, 29, 10928-38	6.6	73
471	Tissue-specific roles of ABCA1 influence susceptibility to atherosclerosis. *Arteriosclerosis, Thrombosis, and Vascular Biology*, **2009**, 29, 548-54	9.4	85
470	Palmitoylation of ATP-binding cassette transporter A1 is essential for its trafficking and function. *Circulation Research*, **2009**, 105, 138-47	15.7	59
469	Despite antiatherogenic metabolic characteristics, SCD1-deficient mice have increased inflammation and atherosclerosis. *Arteriosclerosis, Thrombosis, and Vascular Biology*, **2009**, 29, 341-7	9.4	78
468	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
467	Presynaptic defects underlying impaired learning and memory function in lipoprotein lipase-deficient mice. *Journal of Neuroscience*, **2009**, 29, 4681-5	6.6	43
466	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
465	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
464	Age-dependent alterations of corticostriatal activity in the YAC128 mouse model of Huntington disease. *Journal of Neuroscience*, **2009**, 29, 2414-27	6.6	139
463	Perceptions of genetic discrimination among people at risk for Huntington's disease: a cross sectional survey. *BMJ, The*, **2009**, 338, b2175	5.9	77
462	Neuronal palmitoyl acyl transferases exhibit distinct substrate specificity. *FASEB Journal*, **2009**, 23, 2605-15	6.9	119
461	Application of principal component analysis to pharmacogenomic studies in Canada. *Pharmacogenomics Journal*, **2009**, 9, 362-72	3.5	20
460	Gender differences in expression of the human caspase-12 long variant determines susceptibility to Listeria monocytogenes infection. *Proceedings of the National Academy of Sciences of the United States of America*, **2009**, 106, 9016-20	11.5	51

459	Enhanced susceptibility to pancreatitis in severe hypertriglyceridaemic lipoprotein lipase-deficient mice and agonist-like function of pancreatic lipase in pancreatic cells. *Gut*, **2009**, 58, 422-30	19.2	50
458	IKK phosphorylates Huntingtin and targets it for degradation by the proteasome and lysosome. *Journal of Cell Biology*, **2009**, 187, 1083-99	7.3	287
457	Plasma apolipoprotein AV levels in mice are positively associated with plasma triglyceride levels. *Journal of Lipid Research*, **2009**, 50, 880-4	6.3	13
456	Adverse drug reaction active surveillance: developing a national network in Canada's children's hospitals. *Pharmacoepidemiology and Drug Safety*, **2009**, 18, 713-21	2.6	53
455	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
454	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
453	Balance between synaptic versus extrasynaptic NMDA receptor activity influences inclusions and neurotoxicity of mutant huntingtin. *Nature Medicine*, **2009**, 15, 1407-13	50.5	343
452	The role of free fatty acids, pancreatic lipase and Ca+ signalling in injury of isolated acinar cells and pancreatitis model in lipoprotein lipase-deficient mice. *Acta Physiologica*, **2009**, 195, 13-28	5.6	59
451	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
450	Response to Falush: a role for cis-element polymorphisms in HD. *American Journal of Human Genetics*, **2009**, 85, 942-5	11	4
449	Acute hypertriglyceridemic pancreatitis during pregnancy due to homozygous lipoprotein lipase gene mutation. *Clinica Chimica Acta*, **2009**, 400, 137-8	6.2	6
448	Absence of stearoyl-CoA desaturase-1 does not promote DSS-induced acute colitis. *Biochimica Et Biophysica Acta - Molecular and Cell Biology of Lipids*, **2009**, 1791, 1166-72	5	11
447	Enhanced atherothrombotic formation after oxidative injury by FeCl3 to the common carotid artery in severe combined hyperlipidemic mice. *Biochemical and Biophysical Research Communications*, **2009**, 385, 563-9	3.4	5
446	Phosphorylation of huntingtin reduces the accumulation of its nuclear fragments. *Molecular and Cellular Neurosciences*, **2009**, 40, 121-7	4.8	70
445	Mouse models of Huntington disease: variations on a theme. *DMM Disease Models and Mechanisms*, **2009**, 2, 123-9	4.1	80
444	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
443	Engagement with genetic discrimination: concerns and experiences in the context of Huntington disease. *European Journal of Human Genetics*, **2008**, 16, 279-89	5.3	52
442	The Asn9 variant of lipoprotein lipase is associated with the D9N3G promoter mutation and an increased risk of coronary artery disease. *Clinical Genetics*, **2008**, 53, 27-33	4	

441	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
440	Automated deformation analysis in the YAC128 Huntington disease mouse model. *NeuroImage*, **2008**, 39, 32-9	7.9	136
439	Cortical thickness measured from MRI in the YAC128 mouse model of Huntington's disease. *NeuroImage*, **2008**, 41, 243-51	7.9	98
438	ABCA1 gene mutations, HDL cholesterol levels, and risk of ischemic heart disease. *JAMA - Journal of the American Medical Association*, **2008**, 300, 1997-8; author reply 1998	27.4	12
437	Spontaneous atherosclerosis in aged lipoprotein lipase-deficient mice with severe hypertriglyceridemia on a normal chow diet. *Circulation Research*, **2008**, 102, 250-6	15.7	76
436	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
435	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
434	Response to the Letter by Ebara et al. *Circulation Research*, **2008**, 102,	15.7	1
433	Polyglutamine-modulated striatal calpain activity in YAC transgenic huntington disease mouse model: impact on NMDA receptor function and toxicity. *Journal of Neuroscience*, **2008**, 28, 12725-35	6.6	62
432	Absence of stearoyl-CoA desaturase-1 ameliorates features of the metabolic syndrome in LDLR-deficient mice. *Journal of Lipid Research*, **2008**, 49, 217-29	6.3	51
431	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
430	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
429	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
428	Protective up-regulation of CK2 by mutant huntingtin in cells co-expressing NMDA receptors. *Journal of Neurochemistry*, **2008**, 104, 790-805	6	14
427	Re: Autopsy-proven Huntington's disease with 29 trinucleotide repeats. *Movement Disorders*, **2008**, 23, 1794-5; author reply 1793	7	17
426	Perceptions of discrimination among persons who have undergone predictive testing for Huntington's disease. *American Journal of Medical Genetics Part B: Neuropsychiatric Genetics*, **2008**, 147, 320-5	3.5	37
425	Cholesterol in islet dysfunction and type 2 diabetes. *Journal of Clinical Investigation*, **2008**, 118, 403-8	15.9	105
424	When good drugs go bad. *Nature*, **2007**, 446, 975-7	50.4	188

423	Beta-cell ABCA1 influences insulin secretion, glucose homeostasis and response to thiazolidinedione treatment. *Nature Medicine*, **2007**, 13, 340-7	50.5	315
422	Managing genetic discrimination: strategies used by individuals found to have the Huntington disease mutation. *Clinical Genetics*, **2007**, 71, 220-31	4	29
421	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
420	CAG-encoded polyglutamine length polymorphism in the human genome. *BMC Genomics*, **2007**, 8, 126	4.5	60
419	Phenotypic abnormalities in the YAC128 mouse model of Huntington disease are penetrant on multiple genetic backgrounds and modulated by strain. *Neurobiology of Disease*, **2007**, 26, 189-200	7.5	83
418	Testicular degeneration in Huntington disease. *Neurobiology of Disease*, **2007**, 26, 512-20	7.5	81
417	Verbal episodic memory declines prior to diagnosis in Huntington's disease. *Neuropsychologia*, **2007** , 45, 1767-76	3.2	109
416	Genotypic approaches to therapy in children: a national active surveillance network (GATC) to study the pharmacogenomics of severe adverse drug reactions in children. *Annals of the New York Academy of Sciences*, **2007**, 1110, 177-92	6.5	22
415	Mycophenolate mofetil and atherosclerosis: results of animal and human studies. *Annals of the New York Academy of Sciences*, **2007**, 1110, 209-21	6.5	20
414	Structural abnormalities in spermatids together with reduced sperm counts and motility underlie the reproductive defect in HIP1-/- mice. *Molecular Reproduction and Development*, **2007**, 74, 341-59	2.6	22
413	Altered NMDA receptor trafficking in a yeast artificial chromosome transgenic mouse model of Huntington's disease. *Journal of Neuroscience*, **2007**, 27, 3768-79	6.6	94
412	NMDA receptor function and NMDA receptor-dependent phosphorylation of huntingtin is altered by the endocytic protein HIP1. *Journal of Neuroscience*, **2007**, 27, 2298-308	6.6	37
411	Beyond disgust: impaired recognition of negative emotions prior to diagnosis in Huntington's disease. *Brain*, **2007**, 130, 1732-44	11.2	156
410	N-terminal proteolysis of full-length mutant huntingtin in an inducible PC12 cell model of Huntington's disease. *Cell Cycle*, **2007**, 6, 2970-81	4.7	56
409	Cholesterol biosynthesis pathway is disturbed in YAC128 mice and is modulated by huntingtin mutation. *Human Molecular Genetics*, **2007**, 16, 2187-98	5.6	82
408	Physiologically regulated transgenic ABCA1 does not reduce amyloid burden or amyloid-beta peptide levels in vivo. *Journal of Lipid Research*, **2007**, 48, 914-23	6.3	12
407	Huntingtin-interacting protein 1 influences worm and mouse presynaptic function and protects Caenorhabditis elegans neurons against mutant polyglutamine toxicity. *Journal of Neuroscience*, **2007**, 27, 11056-64	6.6	52
406	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

405	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
404	Expression of LPL in endothelial-intact artery results in lipid deposition and vascular cell adhesion molecule-1 upregulation in both LPL and ApoE-deficient mice. *Arteriosclerosis, Thrombosis, and Vascular Biology*, **2007**, 27, 197-203	9.4	24
403	Glutamate receptor abnormalities in the YAC128 transgenic mouse model of Huntington's disease. *Neuroscience*, **2007**, 147, 354-72	3.9	44
402	Selective degeneration in YAC mouse models of Huntington disease. *Brain Research Bulletin*, **2007**, 72, 124-31	3.9	45
401	Pharmacogenomics and its implications for autoimmune disease. *Journal of Autoimmunity*, **2007**, 28, 122-8	15.5	22
400	Chapter 15 Juvenile amyotrophic lateral sclerosis. *Handbook of Clinical Neurology / Edited By P J Vinken and G W Bruyn*, **2007**, 82, 301-12	3	32
399	To be or not to be toxic: aggregations in Huntington and Alzheimer disease. *Trends in Genetics*, **2006**, 22, 408-11	8.5	29
398	FASA-57 cDNA shares no homology with coding sequence of HD gene. *Journal of Reproductive Immunology*, **2006**, 69, 9-10; author reply 11	4.2	
397	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
396	Levels of mutant huntingtin influence the phenotypic severity of Huntington disease in YAC128 mouse models. *Neurobiology of Disease*, **2006**, 21, 444-55	7.5	73
395	Elevated brain 3-hydroxykynurenine and quinolinate levels in Huntington disease mice. *Neurobiology of Disease*, **2006**, 23, 190-7	7.5	125
394	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
393	Brain-derived neurotrophic factor does not influence age at neurologic onset of Huntington's disease. *Neurobiology of Disease*, **2006**, 24, 280-5	7.5	29
392	Wild-type huntingtin ameliorates striatal neuronal atrophy but does not prevent other abnormalities in the YAC128 mouse model of Huntington disease. *BMC Neuroscience*, **2006**, 7, 80	3.2	43
391	Preparing for preventive clinical trials: the Predict-HD study. *Archives of Neurology*, **2006**, 63, 883-90		259
390	Mycophenolate mofetil (MMF): firing at the atherosclerotic plaque from different angles?. *Cardiovascular Research*, **2006**, 69, 341-7	9.9	34
389	Body weight is modulated by levels of full-length huntingtin. *Human Molecular Genetics*, **2006**, 15, 1513-26	5.6	80
388	Lipoprotein lipase S447X: a naturally occurring gain-of-function mutation. *Arteriosclerosis, Thrombosis, and Vascular Biology*, **2006**, 26, 1236-45	9.4	121

387	Mycophenolate mofetil as an immunomodulatory silver bullet in atherogenesis?. *Lupus*, **2006**, 15, 11-17	2.6	2
386	Mycophenolate mofetil and animal models. *Lupus*, **2006**, 15, 27-34	2.6	
385	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
384	Specific mutations in ABCA1 have discrete effects on ABCA1 function and lipid phenotypes both in vivo and in vitro. *Circulation Research*, **2006**, 99, 389-97	15.7	88
383	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
382	Mutant huntingtin: nuclear translocation and cytotoxicity mediated by GAPDH. *Proceedings of the National Academy of Sciences of the United States of America*, **2006**, 103, 3405-9	11.5	93
381	Hepatic ATP-binding cassette transporter A1 is a key molecule in high-density lipoprotein cholesteryl ester metabolism in mice. *Arteriosclerosis, Thrombosis, and Vascular Biology*, **2006**, 26, 1821-7	9.4	31
380	Both hepatic and extrahepatic ABCA1 have discrete and essential functions in the maintenance of plasma high-density lipoprotein cholesterol levels in vivo. *Circulation*, **2006**, 114, 1301-9	16.7	70
379	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
378	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
377	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
376	Is tetrabenazine safe and effective for suppressing chorea in Huntington's disease?. *Nature Clinical Practice Neurology*, **2006**, 2, 536-7		3
375	Correction of feline lipoprotein lipase deficiency with adeno-associated virus serotype 1-mediated gene transfer of the lipoprotein lipase S447X beneficial mutation. *Human Gene Therapy*, **2006**, 17, 487-99	4.8	63
374	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
373	Hemorheological abnormalities in lipoprotein lipase deficient mice with severe hypertriglyceridemia. *Biochemical and Biophysical Research Communications*, **2006**, 341, 1066-71	3.4	20
372	Hippi is essential for node cilia assembly and Sonic hedgehog signaling. *Developmental Biology*, **2006**, 300, 523-33	3.1	75
371	Molecular Pathogenesis of Huntington's Disease: The Role of Excitotoxicity **2006**, 251-260		1
370	Genome-wide significance for a modifier of age at neurological onset in Huntington's disease at 6q23-24: the HD MAPS study. *BMC Medical Genetics*, **2006**, 7, 71	2.1	62

369	Cerebrospinal fluid levels of orexin-A are not a clinically useful biomarker for Huntington disease. *Clinical Genetics*, **2006**, 70, 78-9	4	29
368	The genotype-phenotype correlation of hereditary multiple exostoses. *Clinical Genetics*, **2006**, 70, 122-30	4	60
367	Predictive testing for Huntington disease: interpretation and significance of intermediate alleles. *Clinical Genetics*, **2006**, 70, 283-94	4	95
366	Palmitoylation of huntingtin by HIP14 is essential for its trafficking and function. *Nature Neuroscience*, **2006**, 9, 824-31	25.5	230
365	Huntingtin inhibits caspase-3 activation. *EMBO Journal*, **2006**, 25, 5896-906	13	73
364	Wild-type huntingtin protects neurons from excitotoxicity. *Journal of Neurochemistry*, **2006**, 96, 1121-9	6	125
363	Intestinal ABCA1 directly contributes to HDL biogenesis in vivo. *Journal of Clinical Investigation*, **2006**, 116, 1052-62	15.9	389
362	Correction of Feline Lipoprotein Lipase Deficiency with Adeno-Associated Virus Serotype 1-Mediated Gene Transfer of the Lipoprotein Lipase S447X Beneficial Mutation. *Human Gene Therapy*, **2006**, 060801084750005	4.8	
361	Gene therapy for lipoprotein lipase deficiency: working toward clinical application. *Human Gene Therapy*, **2005**, 16, 1276-86	4.8	45
360	Cross-species characterization of the ALS2 gene and analysis of its pattern of expression in development and adulthood. *Neurobiology of Disease*, **2005**, 18, 243-57	7.5	24
359	Ethyl-EPA treatment improves motor dysfunction, but not neurodegeneration in the YAC128 mouse model of Huntington disease. *Experimental Neurology*, **2005**, 196, 266-72	5.7	52
358	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
357	Experimental models of Huntington's disease. *Drug Discovery Today: Disease Models*, **2005**, 2, 291-297	1.3	4
356	Cardiovascular disease in systemic lupus erythematosus: has the time for action come?. *Current Opinion in Lipidology*, **2005**, 16, 501-6	4.4	3
355	Treatment of YAC128 mice and their wild-type littermates with cystamine does not lead to its accumulation in plasma or brain: implications for the treatment of Huntington disease. *Journal of Neurochemistry*, **2005**, 94, 1087-101	6	49
354	Cystamine treatment is neuroprotective in the YAC128 mouse model of Huntington disease. *Journal of Neurochemistry*, **2005**, 95, 210-20	6	84
353	Effects of CAG repeat length, HTT protein length and protein context on cerebral metabolism measured using magnetic resonance spectroscopy in transgenic mouse models of Huntington's disease. *Journal of Neurochemistry*, **2005**, 95, 553-62	6	64
352	Introduction to Social and Behavioural Research in Genetics. *Clinical Genetics*, **2005**, 67, 480-480	4	

351	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
350	Ethyl-EPA in Huntington disease: a double-blind, randomized, placebo-controlled trial. *Neurology*, **2005**, 65, 286-92	6.5	125
349	Huntingtin associates with acidic phospholipids at the plasma membrane. *Journal of Biological Chemistry*, **2005**, 280, 36464-73	5.4	117
348	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
347	Complete rescue of lipoprotein lipase-deficient mice by somatic gene transfer of the naturally occurring LPLS447X beneficial mutation. *Arteriosclerosis, Thrombosis, and Vascular Biology*, **2005**, 25, 2143-50	9.4	54
346	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
345	Alternate transcripts expressed in response to diet reflect tissue-specific regulation of ABCA1. *Journal of Lipid Research*, **2005**, 46, 2061-71	6.3	20
344	Complete functional rescue of the ABCA1-/- mouse by human BAC transgenesis. *Journal of Lipid Research*, **2005**, 46, 1113-23	6.3	11
343	Huntingtin interacting protein 1 (HIP1) regulates clathrin assembly through direct binding to the regulatory region of the clathrin light chain. *Journal of Biological Chemistry*, **2005**, 280, 6101-8	5.4	89
342	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
341	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
340	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
339	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
338	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
337	Accurate prediction of the functional significance of single nucleotide polymorphisms and mutations in the ABCA1 gene. *PLoS Genetics*, **2005**, 1, e83	6	107
336	Gene Therapy for Lipoprotein Lipase Deficiency: Working Toward Clinical Application. *Human Gene Therapy*, **2005**, 051014061337001	4.8	1
335	Psychosocial effects of predictive testing for Huntington's disease. *Advances in Neurology*, **2005**, 96, 226-39		11
334	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

333	Deficiency of ABCA1 impairs apolipoprotein E metabolism in brain. *Journal of Biological Chemistry*, **2004**, 279, 41197-207	5.4	263
332	Mutant huntingtin impairs axonal trafficking in mammalian neurons in vivo and in vitro. *Molecular and Cellular Biology*, **2004**, 24, 8195-209	4.8	409
331	The ATP-binding cassette transporter 1 mediates lipid efflux from Sertoli cells and influences male fertility. *Journal of Lipid Research*, **2004**, 45, 1040-50	6.3	70
330	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
329	Huntingtin bodies sequester vesicle-associated proteins by a polyproline-dependent interaction. *Journal of Neuroscience*, **2004**, 24, 269-81	6.6	142
328	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
327	HAP1 facilitates effects of mutant huntingtin on inositol 1,4,5-trisphosphate-induced Ca release in primary culture of striatal medium spiny neurons. *European Journal of Neuroscience*, **2004**, 20, 1779-87	3.5	49
326	A homozygous HAMP mutation in a multiply consanguineous family with pseudo-dominant juvenile hemochromatosis. *Clinical Genetics*, **2004**, 65, 378-83	4	47
325	Mutations in HFE2 cause iron overload in chromosome 1q-linked juvenile hemochromatosis. *Nature Genetics*, **2004**, 36, 77-82	36.3	797
324	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
323	Differential modulation of endotoxin responsiveness by human caspase-12 polymorphisms. *Nature*, **2004**, 429, 75-9	50.4	344
322	Genetic testing and Huntington's disease: issues of employment. *Lancet Neurology, The*, **2004**, 3, 249-52	24.1	22
321	Expression of matrix metalloproteinase activity in idiopathic dilated cardiomyopathy: a marker of cardiac dilatation. *Molecular and Cellular Biochemistry*, **2004**, 264, 183-91	4.2	19
320	Evidence for a modifier of onset age in Huntington disease linked to the HD gene in 4p16. *Neurogenetics*, **2004**, 5, 109-14	3	63
319	HDL deficiency and atherosclerosis: lessons from Tangier disease. *Journal of Internal Medicine*, **2004**, 255, 299-301	10.8	22
318	A case of Tangier disease with a novel mutation in the C-terminal region of ATP-binding cassette transporter A1. *American Journal of Medical Genetics Part A*, **2004**, 130A, 398-401		7
317	Huntington's Disease-like 2 (HDL2) in North America and Japan. *Annals of Neurology*, **2004**, 56, 670-4	9.4	88
316	Long-term correction of murine lipoprotein lipase deficiency with AAV1-mediated gene transfer of the naturally occurring LPL(S447X) beneficial mutation. *Human Gene Therapy*, **2004**, 15, 906-19	4.8	59

315	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
314	Long-term lentiviral-mediated expression of ciliary neurotrophic factor in the striatum of Huntington's disease transgenic mice. *Experimental Neurology*, **2004**, 185, 26-35	5.7	48
313	Deranged neuronal calcium signaling and Huntington disease. *Biochemical and Biophysical Research Communications*, **2004**, 322, 1310-7	3.4	210
312	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
311	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
310	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
309	ABCA1 is essential for efficient basolateral cholesterol efflux during the absorption of dietary cholesterol in chickens. *Journal of Biological Chemistry*, **2003**, 278, 13356-66	5.4	92
308	Alterations of plasma lipids in mice via adenoviral-mediated hepatic overexpression of human ABCA1. *Journal of Lipid Research*, **2003**, 44, 1470-80	6.3	74
307	A role for epsin N-terminal homology/AP180 N-terminal homology (ENTH/ANTH) domains in tubulin binding. *Journal of Biological Chemistry*, **2003**, 278, 28823-30	5.4	24
306	Nuclear localization of a non-caspase truncation product of atrophin-1, with an expanded polyglutamine repeat, increases cellular toxicity. *Journal of Biological Chemistry*, **2003**, 278, 13047-55	5.4	68
305	Selective striatal neuronal loss in a YAC128 mouse model of Huntington disease. *Human Molecular Genetics*, **2003**, 12, 1555-67	5.6	609
304	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
303	The role of the ABCA1 transporter and cholesterol efflux in familial hypoalphalipoproteinemia. *Journal of Lipid Research*, **2003**, 44, 1251-5	6.3	35
302	Mutation screening of the ALS2 gene in sporadic and familial amyotrophic lateral sclerosis. *Archives of Neurology*, **2003**, 60, 1768-71		36
301	Disruption of the endocytic protein HIP1 results in neurological deficits and decreased AMPA receptor trafficking. *EMBO Journal*, **2003**, 22, 3254-66	13	91
300	Predictive testing for Huntington's disease: a universal model?. *Lancet Neurology, The*, **2003**, 2, 141-2	24.1	26
299	Huntingtin proteolysis in Huntington disease. *Clinical Neuroscience Research*, **2003**, 3, 129-139		9
298	Interaction of normal and expanded CAG repeat sizes influences age at onset of Huntington disease **2003**, 119A, 279-82		117

297	CAG repeat polymorphisms in KCNN3 (HSKCa3) and PPP2R2B show no association or linkage to schizophrenia **2003**, 116B, 45-50		9
296	An ALS2 gene mutation causes hereditary spastic paraplegia in a Pakistani kindred. *Annals of Neurology*, **2003**, 53, 144-5	9.4	81
295	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
294	The first nonsense mutation in alsin results in a homogeneous phenotype of infantile-onset ascending spastic paralysis with bulbar involvement in two siblings. *Clinical Genetics*, **2003**, 64, 210-5	4	70
293	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
292	Predictive testing for persons at risk for homozygosity for CAG expansion in the Huntington disease gene. *Clinical Genetics*, **2003**, 64, 524-5	4	9
291	Depletion of wild-type huntingtin in mouse models of neurologic diseases. *Journal of Neurochemistry*, **2003**, 87, 101-6	6	81
290	Huntingtin interacts with REST/NRSF to modulate the transcription of NRSE-controlled neuronal genes. *Nature Genetics*, **2003**, 35, 76-83	36.3	709
289	Clarity is essential when using nucleotide number systems. *Atherosclerosis*, **2003**, 170, 349	3.1	
288	Role of NR2B-type NMDA receptors in selective neurodegeneration in Huntington disease. *Neurobiology of Aging*, **2003**, 24, 1113-21	5.6	92
287	Caudate volume as an outcome measure in clinical trials for Huntington's disease: a pilot study. *Brain Research Bulletin*, **2003**, 62, 137-41	3.9	37
286	Heterozygosity for ABCA1 gene mutations: effects on enzymes, apolipoproteins and lipoprotein particle size. *Atherosclerosis*, **2003**, 171, 311-9	3.1	17
285	Skewed X-chromosome inactivation is associated with trisomy in women ascertained on the basis of recurrent spontaneous abortion or chromosomally abnormal pregnancies. *American Journal of Human Genetics*, **2003**, 72, 399-407	11	68
284	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
283	Allelic variation in the promoter region of the LDL receptor gene: analysis of an African-specific variant in the FP2 cis-acting regulatory element. *Molecular and Cellular Probes*, **2003**, 17, 175-81	3.3	1
282	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
281	Homozygosity for CAG mutation in Huntington disease is associated with a more severe clinical course. *Brain*, **2003**, 126, 946-55	11.2	151
280	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

279	Caspase cleavage of mutant huntingtin precedes neurodegeneration in Huntington's disease. *Journal of Neuroscience*, **2002**, 22, 7862-72	6.6	293
278	Striatal neurochemical changes in transgenic models of Huntington's disease. *Journal of Neuroscience Research*, **2002**, 68, 716-29	4.4	83
277	ABCA1 regulatory variants influence coronary artery disease independent of effects on plasma lipid levels. *Clinical Genetics*, **2002**, 61, 115-25	4	85
276	ABCA1 mRNA and protein distribution patterns predict multiple different roles and levels of regulation. *Laboratory Investigation*, **2002**, 82, 273-83	5.9	239
275	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
274	Mutant DNA-binding domain of HSF4 is associated with autosomal dominant lamellar and Marner cataract. *Nature Genetics*, **2002**, 31, 276-8	36.3	215
273	Mutant frizzled-4 disrupts retinal angiogenesis in familial exudative vitreoretinopathy. *Nature Genetics*, **2002**, 32, 326-30	36.3	368
272	Early mitochondrial calcium defects in Huntington's disease are a direct effect of polyglutamines. *Nature Neuroscience*, **2002**, 5, 731-6	25.5	839
271	Targeted disruption of Huntingtin-associated protein-1 (Hap1) results in postnatal death due to depressed feeding behavior. *Human Molecular Genetics*, **2002**, 11, 945-59	5.6	66
270	HIP14, a novel ankyrin domain-containing protein, links huntingtin to intracellular trafficking and endocytosis. *Human Molecular Genetics*, **2002**, 11, 2815-28	5.6	156
269	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
268	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
267	Identification and functional analysis of a naturally occurring E89K mutation in the ABCA1 gene of the WHAM chicken. *Journal of Lipid Research*, **2002**, 43, 1610-7	6.3	38
266	HIP1 and HIP12 display differential binding to F-actin, AP2, and clathrin. Identification of a novel interaction with clathrin light chain. *Journal of Biological Chemistry*, **2002**, 277, 19897-904	5.4	84
265	Truncation mutations in ABCA1 suppress normal upregulation of full-length ABCA1 by 9-cis-retinoic acid and 22-R-hydroxycholesterol. *Journal of Lipid Research*, **2002**, 43, 1939-49	6.3	25
264	Expression and functional analyses of novel mutations of ATP-binding cassette transporter-1 in Japanese patients with high-density lipoprotein deficiency. *Biochemical and Biophysical Research Communications*, **2002**, 290, 713-21	3.4	29
263	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
262	Association between increased arterial-wall thickness and impairment in ABCA1-driven cholesterol efflux: an observational study. *Lancet, The*, **2002**, 359, 37-42	40	171

261	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
260	Increased ABCA1 activity protects against atherosclerosis. *Journal of Clinical Investigation*, **2002**, 110, 35-42	15.9	203
259	Increased ABCA1 activity protects against atherosclerosis. *Journal of Clinical Investigation*, **2002**, 110, 35-42	15.9	95
258	Structural and functional consequences of missense mutations in exon 5 of the lipoprotein lipase gene. *Journal of Lipid Research*, **2002**, 43, 398-406	6.3	25
257	High incidence rate and absent family histories in one quarter of patients newly diagnosed with Huntington disease in British Columbia. *Clinical Genetics*, **2001**, 60, 198-205	4	73
256	The "flap" endonuclease gene FEN1 is excluded as a candidate gene implicated in the CAG repeat expansion underlying Huntington disease. *Clinical Genetics*, **2001**, 59, 122-7	4	31
255	The LPL S447X cSNP is associated with decreased blood pressure and plasma triglycerides, and reduced risk of coronary artery disease. *Clinical Genetics*, **2001**, 60, 293-300	4	41
254	NMDA receptor function in mouse models of Huntington disease. *Journal of Neuroscience Research*, **2001**, 66, 525-39	4.4	218
253	Familial influence on age of onset among siblings with Huntington disease*. *American Journal of Medical Genetics Part A*, **2001**, 105, 399-403		75
252	addendum: A one-hit model of cell death in inherited neuronal degenerations. *Nature*, **2001**, 409, 542-543	30.4	2
251	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
250	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
249	Double-stranded RNA-dependent protein kinase, PKR, binds preferentially to Huntington's disease (HD) transcripts and is activated in HD tissue. *Human Molecular Genetics*, **2001**, 10, 1531-8	5.6	78
248	Clinical markers of early disease in persons near onset of Huntington's disease. *Neurology*, **2001**, 57, 658-62	6.3	180
247	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
246	Maternal expression of functional lipoprotein lipase and effects on body fat mass and body condition scores of mature cats with lipoprotein lipase deficiency. *American Journal of Veterinary Research*, **2001**, 62, 264-9	1.1	9
245	The factor structure for positive and negative symptoms in South African Xhosa patients with schizophrenia. *Schizophrenia Research*, **2001**, 47, 149-57	3.6	36
244	Common mutations in the lipoprotein lipase gene (LPL): effects on HDL-cholesterol levels in a Chinese Canadian population. *Atherosclerosis*, **2001**, 156, 401-7	3.1	29

243	Loss of huntingtin-mediated BDNF gene transcription in Huntington's disease. *Science*, **2001**, 293, 493-8	33.3	1044
242	Cloning and characterization of three novel genes, ALS2CR1, ALS2CR2, and ALS2CR3, in the juvenile amyotrophic lateral sclerosis (ALS2) critical region at chromosome 2q33-q34: candidate genes for ALS2. *Genomics*, **2001**, 71, 200-13	4.3	43
241	Mutant huntingtin enhances excitotoxic cell death. *Molecular and Cellular Neurosciences*, **2001**, 17, 41-53	4.8	158
240	A gene encoding a putative GTPase regulator is mutated in familial amyotrophic lateral sclerosis 2. *Nature Genetics*, **2001**, 29, 166-73	36.3	552
239	Measurement of mutational flow implies both a high new-mutation rate for Huntington disease and substantial underascertainment of late-onset cases. *American Journal of Human Genetics*, **2001**, 68, 373-85	11	55
238	Wild-type huntingtin reduces the cellular toxicity of mutant huntingtin in vivo. *American Journal of Human Genetics*, **2001**, 68, 313-24	11	183
237	Onset and pre-onset studies to define the Huntington's disease natural history. *Brain Research Bulletin*, **2001**, 56, 233-8	3.9	41
236	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
235	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
234	The Gln-Ala repeat transcriptional activator CA150 interacts with huntingtin: neuropathologic and genetic evidence for a role in Huntington's disease pathogenesis. *Proceedings of the National Academy of Sciences of the United States of America*, **2001**, 98, 1811-6	11.5	81
233	Cholesterol efflux regulatory protein, Tangier disease and familial high-density lipoprotein deficiency. *Current Opinion in Lipidology*, **2000**, 11, 117-22	4.4	99
232	Caspases and neurodegeneration: on the cutting edge of new therapeutic approaches. *Clinical Genetics*, **2000**, 57, 1-10	4	106
231	A one-hit model of cell death in inherited neuronal degenerations. *Nature*, **2000**, 406, 195-9	50.4	250
230	Michael Smith (1932-2000). *Nature*, **2000**, 408, 786	50.4	
229	HIP12 is a non-proapoptotic member of a gene family including HIP1, an interacting protein with huntingtin. *Mammalian Genome*, **2000**, 11, 1006-15	3.2	40
228	Wild-type huntingtin protects from apoptosis upstream of caspase-3. *Journal of Neuroscience*, **2000**, 20, 3705-13	6.6	310
227	Huntingtin interacting protein 1 induces apoptosis via a novel caspase-dependent death effector domain. *Journal of Biological Chemistry*, **2000**, 275, 41299-308	5.4	94
226	Phenotypic correction of feline lipoprotein lipase deficiency by adenoviral gene transfer. *Human Gene Therapy*, **2000**, 11, 21-32	4.8	36

225	Huntingtin is required for normal hematopoiesis. *Human Molecular Genetics*, **2000**, 9, 387-94	5.6	31
224	Familial aggregation of psychotic symptoms in Huntington's disease. *American Journal of Psychiatry*, **2000**, 157, 1955-9	11.9	50
223	Lipoprotein lipase activity is associated with severity of angina pectoris. REGRESS Study Group. *Circulation*, **2000**, 102, 1629-33	16.7	45
222	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
221	Human huntingtin-associated protein (HAP-1) gene: genomic organisation and an intragenic polymorphism. *Gene*, **2000**, 254, 181-7	3.8	4
220	Predictive testing for Huntington's disease: the calm after the storm. *Lancet, The*, **2000**, 356, 1944-5	40	40
219	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
218	Huntington disease: new insights on the role of huntingtin cleavage. *Journal of Neural Transmission Supplementum*, **2000**, 1-17		21
217	Plasma and vessel wall lipoprotein lipase have different roles in atherosclerosis. *Journal of Lipid Research*, **2000**, 41, 521-531	6.3	67
216	Subtype-specific enhancement of NMDA receptor currents by mutant huntingtin. *Journal of Neurochemistry*, **1999**, 72, 1890-8	6	150
215	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
214	Recent insights into the molecular pathogenesis of Huntington disease. *Seminars in Neurology*, **1999**, 19, 385-95	3.2	40
213	Cleavage of atrophin-1 at caspase site aspartic acid 109 modulates cytotoxicity. *Journal of Biological Chemistry*, **1999**, 274, 8730-6	5.4	90
212	A frequent mutation in the lipoprotein lipase gene (D9N) deteriorates the biochemical and clinical phenotype of familial hypercholesterolemia. *Arteriosclerosis, Thrombosis, and Vascular Biology*, **1999**, 19, 2708-13	9.4	27
211	In vitro evidence for both the nucleus and cytoplasm as subcellular sites of pathogenesis in Huntington's disease. *Human Molecular Genetics*, **1999**, 8, 25-33	5.6	69
210	Accurate determination of the number of CAG repeats in the Huntington disease gene using a sequence-specific internal DNA standard. *Clinical Genetics*, **1999**, 55, 198-202	4	24
209	A common truncation variant of lipoprotein lipase (Ser447X) confers protection against coronary heart disease: the Framingham Offspring Study. *Clinical Genetics*, **1999**, 55, 450-4	4	92
208	Two common mutations (D9N, N291S) in lipoprotein lipase: a cumulative analysis of their influence on plasma lipids and lipoproteins in men and women. *Clinical Genetics*, **1999**, 56, 297-305	4	36

207	Lipid and lipoprotein analysis of cats with lipoprotein lipase deficiency. *European Journal of Clinical Investigation*, **1999**, 29, 17-26	4.6	29
206	Kennedy's disease: caspase cleavage of the androgen receptor is a crucial event in cytotoxicity. *Journal of Neurochemistry*, **1999**, 72, 185-95	6	173
205	Life without huntingtin: normal differentiation into functional neurons. *Journal of Neurochemistry*, **1999**, 72, 1009-18	6	33
204	Mutations in ABC1 in Tangier disease and familial high-density lipoprotein deficiency. *Nature Genetics*, **1999**, 22, 336-45	36.3	1468
203	Chromosomal localization of the Huntingtin associated protein (HAP-1) gene in mouse and humans with radiation hybrid and interspecific backcross mapping. *Mammalian Genome*, **1999**, 10, 397-8	3.2	5
202	Genomic organization of the human caspase-9 gene on Chromosome 1p36. 1-p36.3. *Mammalian Genome*, **1999**, 10, 757-60	3.2	12
201	Neuronal degeneration in the basal ganglia and loss of pallido-subthalamic synapses in mice with targeted disruption of the Huntington's disease gene. *Brain Research*, **1999**, 818, 468-79	3.7	65
200	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
199	Forskolin and dopamine D1 receptor activation increase huntingtin's association with endosomes in immortalized neuronal cells of striatal origin. *Neuroscience*, **1999**, 89, 1159-67	3.9	43
198	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
197	Mutations in the ABC1 gene in familial HDL deficiency with defective cholesterol efflux. *Lancet, The* , **1999**, 354, 1341-6	40	321
196	Evidence for both the nucleus and cytoplasm as subcellular sites of pathogenesis in Huntington's disease in cell culture and in transgenic mice expressing mutant huntingtin. *Philosophical Transactions of the Royal Society B: Biological Sciences*, **1999**, 354, 1047-55	5.8	21
195	A yeast artificial chromosome-based physical map of the juvenile amyotrophic lateral sclerosis (ALS2) critical region on human chromosome 2q33-q34. *Genomics*, **1999**, 55, 106-12	4.3	18
194	Lipoprotein lipase activity is decreased in a large cohort of patients with coronary artery disease and is associated with changes in lipids and lipoproteins. *Journal of Lipid Research*, **1999**, 40, 735-743	6.3	68
193	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
192	Length of huntingtin and its polyglutamine tract influences localization and frequency of intracellular aggregates. *Nature Genetics*, **1998**, 18, 150-4	36.3	423
191	Compound heterozygosity for frameshift mutations in the gene for lipoprotein lipase in a patient with early-onset chylomicronemia. *Human Mutation*, **1998**, Suppl 1, S141-4	4.7	4
190	Dyslipidemias associated with heterozygous lipoprotein lipase mutations in the French-Canadian population. *Human Mutation*, **1998**, Suppl 1, S148-53	4.7	11

189	Ile225Thr loop mutation in the lipoprotein lipase (LPL) gene is a de novo event. *American Journal of Medical Genetics Part A*, **1998**, 78, 313-6		10
188	Gene structure and map location of the murine homolog of the Huntington-associated protein, Hap1. *Mammalian Genome*, **1998**, 9, 565-70	3.2	21
187	The new Clinical Genetics: towards the millennium. *Clinical Genetics*, **1998**, 53, 1-2	4	
186	A novel Glu421Lys substitution in the lipoprotein lipase gene in pregnancy-induced hypertriglyceridemic pancreatitis. *Clinica Chimica Acta*, **1998**, 269, 1-12	6.2	35
185	A Huntington disease-like neurodegenerative disorder maps to chromosome 20p. *American Journal of Human Genetics*, **1998**, 63, 1431-8	11	55
184	The fatal attraction of polyglutamine-containing proteins. *Clinical Genetics*, **1998**, 53, 233-42	4	25
183	The influence of huntingtin protein size on nuclear localization and cellular toxicity. *Journal of Cell Biology*, **1998**, 141, 1097-105	7.3	280
182	A common mutation in the lipoprotein lipase gene (N291S) alters the lipoprotein phenotype and risk for cardiovascular disease in patients with familial hypercholesterolemia. *Circulation*, **1998**, 97, 729-35	16.7	72
181	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
180	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
179	The Asn9 variant of lipoprotein lipase is associated with the -93G promoter mutation and an increased risk of coronary artery disease. The Regress Study Group. *Clinical Genetics*, **1998**, 53, 27-33	4	38
178	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
177	Pathological association and dissociation of functional systems in multiple sclerosis and Huntington's disease. *Journal of Clinical and Experimental Neuropsychology*, **1997**, 19, 63-76	2.1	5
176	Assessment of French patients with LPL deficiency for French Canadian mutations. *Journal of Medical Genetics*, **1997**, 34, 672-5	5.8	9
175	Segments in the C-terminal folding domain of lipoprotein lipase important for binding to the low density lipoprotein receptor-related protein and to heparan sulfate proteoglycans. *Journal of Biological Chemistry*, **1997**, 272, 5821-7	5.4	40
174	Rethinking genotype and phenotype correlations in polyglutamine expansion disorders. *Human Molecular Genetics*, **1997**, 6, 2005-10	5.6	116
173	Of molecular interactions, mice and mechanisms: new insights into Huntington's disease. *Current Opinion in Neurology*, **1997**, 10, 291-8	7.1	36
172	Efficient adenovirus-mediated ectopic gene expression of human lipoprotein lipase in human hepatic (HepG2) cells. *Human Gene Therapy*, **1997**, 8, 205-14	4.8	10

171	Familial defective apolipoprotein B-100 in hypercholesterolemic Chinese Canadians: identification of a unique haplotype of the apolipoprotein B-100 allele. *Atherosclerosis*, **1997**, 135, 181-5	3.1	4
170	Risk reversals in predictive testing for Huntington disease. *American Journal of Human Genetics*, **1997**, 61, 945-52	11	28
169	Ethnic variation and in vivo effects of the -93t-->g promoter variant in the lipoprotein lipase gene. *Arteriosclerosis, Thrombosis, and Vascular Biology*, **1997**, 17, 2672-8	9.4	38
168	Correction of hypertriglyceridemia and impaired fat tolerance in lipoprotein lipase-deficient mice by adenovirus-mediated expression of human lipoprotein lipase. *Arteriosclerosis, Thrombosis, and Vascular Biology*, **1997**, 17, 2532-9	9.4	53
167	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
166	Toward understanding the molecular pathology of Huntington's disease. *Brain Pathology*, **1997**, 7, 979-1002		68
165	Localization of the cell death genes CPP32 and Mch-2 to human chromosome 4q. *Mammalian Genome*, **1997**, 8, 56-9	3.2	12
164	Interleukin-1beta-converting enzyme (ICE) and related cell death genes ICErel-II and ICErel-III map to the same PAC clone at band 11q22.2-22.3. *Mammalian Genome*, **1997**, 8, 611-3	3.2	10
163	Dilemmas of anonymous predictive testing for Huntington disease: Privacy vs. optimal care **1997**, 71, 197-201		16
162	A single Ser259Arg mutation in the gene for lipoprotein lipase causes chylomicronemia in Moroccans of Berber ancestry. *Human Mutation*, **1997**, 10, 179-85	4.7	6
161	Differences in the phenotype between children with familial defective apolipoprotein B-100 and familial hypercholesterolemia. *Arteriosclerosis, Thrombosis, and Vascular Biology*, **1997**, 17, 826-33	9.4	23
160	Genetic variant showing a positive interaction with beta-blocking agents with a beneficial influence on lipoprotein lipase activity, HDL cholesterol, and triglyceride levels in coronary artery disease patients. The Ser447-stop substitution in the lipoprotein lipase gene. REGRESS Study Group. *Circulation*, **1997**, 95, 2628-35	16.7	94
159	Human huntingtin derived from YAC transgenes compensates for loss of murine huntingtin by rescue of the embryonic lethal phenotype. *Human Molecular Genetics*, **1996**, 5, 1875-85	5.6	77
158	Gene-based therapeutic strategies for Human Lipoprotein Lipase (LPL) deficiency: Rationale and prospects for alteration of atherogenic risk. *Transfusion Science*, **1996**, 17, 79-87		
157	Huntington disease: new insights into the relationship between CAG expansion and disease. *Human Molecular Genetics*, **1996**, 5 Spec No, 1431-5	5.6	56
156	Compound heterozygosity for a known and a novel defect in the lipoprotein lipase gene (Asp250-->Asn; Ser251-->Cys) resulting in lipoprotein lipase (LPL) deficiency. *Netherlands Journal of Medicine*, **1996**, 49, 189-95	0.5	13
155	Common sequence variants of lipoprotein lipase: standardized studies of in vitro expression and catalytic function. *Lipids and Lipid Metabolism*, **1996**, 1302, 159-66		81
154	A new mutation destroying disulphide bridging in the C-terminal domain of lipoprotein lipase. *Biochemical and Biophysical Research Communications*, **1996**, 227, 189-94	3.4	26

153	Auxiliary-directed peroxidation of 1,4-dienes. *Tetrahedron*, **1996**, 52, 12381-12398	2.4	7
152	Cleavage of huntingtin by apopain, a proapoptotic cysteine protease, is modulated by the polyglutamine tract. *Nature Genetics*, **1996**, 13, 442-9	36.3	501
151	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
150	Adverse psychological events occurring in the first year after predictive testing for Huntington's disease. The Canadian Collaborative Study Predictive Testing. *Journal of Medical Genetics*, **1996**, 33, 856-62	5.8	46
149	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
148	Huntingtin is ubiquitinated and interacts with a specific ubiquitin-conjugating enzyme. *Journal of Biological Chemistry*, **1996**, 271, 19385-94	5.4	263
147	The Asp9 Asn mutation in the lipoprotein lipase gene is associated with increased progression of coronary atherosclerosis. REGRESS Study Group, Interuniversity Cardiology Institute, Utrecht, The Netherlands. Regression Growth Evaluation Statin Study. *Circulation*, **1996**, 94, 1913-8	16.7	78
146	A mutation in the lipoprotein lipase gene is the molecular basis of chylomicronemia in a colony of domestic cats. *Journal of Clinical Investigation*, **1996**, 97, 1257-66	15.9	59
145	Mapping of the gonadotropin-releasing hormone (GnRH) receptor gene to human chromosome 4q21.2 by fluorescence in situ hybridization. *Mammalian Genome*, **1995**, 6, 309-10	3.2	9
144	A lipoprotein lipase mutation (Asn291Ser) is associated with reduced HDL cholesterol levels in premature atherosclerosis. *Nature Genetics*, **1995**, 10, 28-34	36.3	213
143	Many roads lead to atheroma. *Nature Medicine*, **1995**, 1, 22-3	50.5	19
142	Mutations in the gene for lipoprotein lipase. A cause for low HDL cholesterol levels in individuals heterozygous for familial hypercholesterolemia. *Arteriosclerosis, Thrombosis, and Vascular Biology*, **1995**, 15, 1704-12	9.4	38
141	Patients with apoE3 deficiency (E2/2, E3/2, and E4/2) who manifest with hyperlipidemia have increased frequency of an Asn 291-->Ser mutation in the human LPL gene. *Arteriosclerosis, Thrombosis, and Vascular Biology*, **1995**, 15, 1695-703	9.4	57
140	Retroviral-mediated gene transfer and expression of human lipoprotein lipase in somatic cells. *Human Gene Therapy*, **1995**, 6, 853-63	4.8	7
139	Ancestral differences in the distribution of the delta 2642 glutamic acid polymorphism is associated with varying CAG repeat lengths on normal chromosomes: insights into the genetic evolution of Huntington disease. *Human Molecular Genetics*, **1995**, 4, 207-14	5.6	67
138	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
137	Somatic mosaicism in sperm is associated with intergenerational (CAG)n changes in Huntington disease. *Human Molecular Genetics*, **1995**, 4, 189-95	5.6	64
136	A frequently occurring mutation in the lipoprotein lipase gene (Asn291Ser) contributes to the expression of familial combined hyperlipidemia. *Human Molecular Genetics*, **1995**, 4, 1543-9	5.6	74

135	Origins and evolution of Huntington disease chromosomes. *Experimental Neurology*, **1995**, 4, 239-44		21
134	Structural analysis of the 5' region of mouse and human Huntington disease genes reveals conservation of putative promoter region and di- and trinucleotide polymorphisms. *Genomics*, **1995**, 25, 707-15	4.3	35
133	Genomic organization of the human alpha-adducin gene and its alternately spliced isoforms. *Genomics*, **1995**, 25, 93-9	4.3	32
132	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
131	Long-term efficacy and tolerability of simvastatin in a large cohort of elderly hypercholesterolemic patients. *Atherosclerosis*, **1995**, 116, 153-62	3.1	14
130	Hereditary late-onset chorea without significant dementia: genetic evidence for substantial phenotypic variation in Huntington's disease. *Neurology*, **1995**, 45, 443-7	6.5	48
129	Are we all of one mind? Clinicians' and patients' opinions regarding the development of a service protocol for predictive testing for Huntington disease. Canadian Collaborative Study for Predictive Testing for Huntington Disease. *American Journal of Medical Genetics Part A*, **1995**, 58, 59-69		20
128	Gene environment interaction and plasma triglyceride levels: the crucial role of lipoprotein lipase. *Clinical Genetics*, **1994**, 46, 15-8	4	12
127	Sequence of the murine Huntington disease gene: evidence for conservation, alternate splicing and polymorphism in a triplet (CCG) repeat [corrected]. *Human Molecular Genetics*, **1994**, 3, 85-92	5.6	80
126	Analysis of DNA changes in the LPL gene in patients with familial combined hyperlipidemia. *Arteriosclerosis and Thrombosis: A Journal of Vascular Biology*, **1994**, 14, 1250-7		66
125	Apolipoprotein CII-Padova (Tyr37-->stop) as a cause of chylomicronaemia in an Italian kindred from Siculiana. *Journal of Medical Genetics*, **1994**, 31, 622-6	5.8	6
124	Homozygosity for a mutation in the lipoprotein lipase gene (Gly139-->Ser) causes chylomicronaemia in a boy of Spanish descent. *Human Genetics*, **1994**, 93, 339-43	6.3	9
123	Recurrent missense mutations at the first and second base of codon Arg243 in human lipoprotein lipase in patients of different ancestries. *Human Mutation*, **1994**, 3, 52-8	4.7	23
122	Regional cerebral glucose metabolism in huntington's disease: A statistical investigation. *Human Brain Mapping*, **1994**, 2, 95-102	5.9	1
121	U-type exchange in a paracentric inversion as a possible mechanism of origin of an inverted tandem duplication of chromosome 8. *American Journal of Medical Genetics Part A*, **1994**, 49, 384-7		30
120	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
119	Comparative efficacy and safety of pravastatin, nicotinic acid and the two combined in patients with hypercholesterolemia. *American Journal of Cardiology*, **1994**, 73, 339-45	3	73
118	Somatic and gonadal mosaicism of the Huntington disease gene CAG repeat in brain and sperm. *Nature Genetics*, **1994**, 6, 409-14	36.3	331

117	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
116	Precise mapping of the brain alpha 2-adrenergic receptor gene within chromosome 4p16. *Genomics*, **1994**, 19, 298-302	4.3	22
115	The murine homologues of the Huntington disease gene (Hdh) and the alpha-adducin gene (Add1) map to mouse chromosome 5 within a region of conserved synteny with human chromosome 4p16.3. *Genomics*, **1994**, 22, 198-201	4.3	16
114	Murine alpha-L-iduronidase: cDNA isolation and expression. *Genomics*, **1994**, 24, 311-6	4.3	29
113	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
112	Issues in molecular genetic testing of individuals with suspected early-onset familial Alzheimer's disease. *Alzheimer Disease and Associated Disorders*, **1994**, 8, 116-25	2.5	10
111	The molecular genetics of Huntington's disease. *Current Opinion in Neurology*, **1994**, 7, 325-32	7.1	19
110	Normal CAG repeat length in the Huntington's disease gene in senile chorea. *Neurology*, **1994**, 44, 2183-4	6.5	20
109	On planting alfalfa and growing orchids: the cloning of the gene causing Huntington disease. *Clinical Genetics*, **1993**, 43, 217-22	4	2
108	DNA analysis of distinct populations suggests multiple origins for the mutation causing Huntington disease. *Clinical Genetics*, **1993**, 43, 286-94	4	20
107	Mapping of the human NMDA receptor subunit (NMDAR1) and the proposed NMDA receptor glutamate-binding subunit (NMDARA1) to chromosomes 9q34.3 and chromosome 8, respectively. *Genomics*, **1993**, 17, 237-9	4.3	14
106	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
105	A PCR method for accurate assessment of trinucleotide repeat expansion in Huntington disease. *Human Molecular Genetics*, **1993**, 2, 635-6	5.6	64
104	Prevention of raised low-density lipoprotein cholesterol in a patient with familial hypercholesterolaemia and lipoprotein lipase deficiency. *Lancet, The*, **1993**, 341, 1119-21	40	30
103	Monoamines and their metabolites in Huntington's disease brain: evidence for decreased catechol-O-methyltransferase activity. *Biological Psychiatry*, **1993**, 33, 551-3	7.9	4
102	(CA)n-dinucleotide repeat at the PDEB locus in 4p16.3. *Human Molecular Genetics*, **1993**, 2, 827	5.6	7
101	A transcription map of the region containing the Huntington disease gene. *Human Molecular Genetics*, **1993**, 2, 901-7	5.6	40
100	An Alu element retroposition in two families with Huntington disease defines a new active Alu subfamily. *Nucleic Acids Research*, **1993**, 21, 3379-83	20.1	44

99	Five year study of prenatal testing for Huntington's disease: demand, attitudes, and psychological assessment. *Journal of Medical Genetics*, **1993**, 30, 549-56	5.8	70
98	George Huntington: the man behind the eponym. *Journal of Medical Genetics*, **1993**, 30, 406-9	5.8	8
97	Differential 3' polyadenylation of the Huntington disease gene results in two mRNA species with variable tissue expression. *Human Molecular Genetics*, **1993**, 2, 1541-5	5.6	89
96	Molecular analysis of late onset Huntington's disease. *Journal of Medical Genetics*, **1993**, 30, 991-5	5.8	46
95	A 4 basepair deletion in exon 4 of the human lipoprotein lipase gene results in type I hyperlipoproteinemia. *Human Molecular Genetics*, **1993**, 2, 1049-50	5.6	4
94	Familial predisposition to recurrent mutations causing Huntington's disease: genetic risk to sibs of sporadic cases. *Journal of Medical Genetics*, **1993**, 30, 987-90	5.8	27
93	Phenotypic variation of mutations in the human lipoprotein-lipase gene. *Biochemical Society Transactions*, **1993**, 21, 506-9	5.1	17
92	Reply to Dr. Kessler. *American Journal of Medical Genetics Part A*, **1993**, 45, 696-697		1
91	Diagnosis of Huntington disease: a model for the stages of psychological response based on experience of a predictive testing program. *American Journal of Medical Genetics Part A*, **1993**, 47, 368-74		25
90	South African founder mutations in the low-density lipoprotein receptor gene causing familial hypercholesterolemia in the Dutch population. *Human Genetics*, **1993**, 92, 567-70	6.3	22
89	The relationship between trinucleotide (CAG) repeat length and clinical features of Huntington's disease. *Nature Genetics*, **1993**, 4, 398-403	36.3	868
88	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
87	Identification of an Alu retrotransposition event in close proximity to a strong candidate gene for Huntington's disease. *Nature*, **1993**, 362, 370-3	50.4	48
86	Development of a program for identification of patients with familial hypercholesterolemia in British Columbia: a model for prevention of coronary disease. *American Journal of Cardiology*, **1993**, 72, 25D-29D	3	4
85	Acidic and basic fibroblast growth factor-like immunoreactivity in the striatum and midbrain in Huntington's disease. *Brain Research*, **1993**, 610, 1-7	3.7	31
84	Genetic and phenotypic heterogeneity in familial lecithin: cholesterol acyltransferase (LCAT) deficiency. Six newly identified defective alleles further contribute to the structural heterogeneity in this disease. *Journal of Clinical Investigation*, **1993**, 91, 677-83	15.9	72
83	Gene-environment interaction in the conversion of a mild-to-severe phenotype in a patient homozygous for a Ser172-->Cys mutation in the lipoprotein lipase gene. *Journal of Clinical Investigation*, **1993**, 91, 1953-8	15.9	48
82	Recurrent pancreatitis and chylomicronemia in an extended Dutch kindred is caused by a Gly154Ser substitution in lipoprotein lipase.. *Journal of Lipid Research*, **1993**, 34, 2109-2119	6.3	24

81	Molecular Genetics of Lipoprotein Lipase Deficiency. *Medical Science Symposia Series*, **1993**, 97-100		
80	Familial defective apolipoprotein B-100 is clinically indistinguishable from familial hypercholesterolemia. *Archives of Internal Medicine*, **1993**, 153, 2349-2356		18
79	Attitudes toward direct predictive testing for the Huntington disease gene. Relevance for other adult-onset disorders. The Canadian Collaborative Group on Predictive Testing for Huntington Disease. *JAMA - Journal of the American Medical Association*, **1993**, 270, 2321-2325	27.4	42
78	Prevalence, geographical distribution and genealogical investigations of mutation 188 of lipoprotein lipase gene in the French Canadian population of QuÞbec. *Clinical Genetics*, **1992**, 41, 206-10	4	23
77	The prediction of exons through an analysis of spliceable open reading frames. *Nucleic Acids Research*, **1992**, 20, 3453-62	20.1	64
76	The lipoprotein lipase Gly188----Glu mutation in South Africans of Indian descent: evidence suggesting common origins and an increased frequency. *Journal of Medical Genetics*, **1992**, 29, 119-22	5.8	10
75	(CA)n-dinucleotide repeat polymorphism at the locus for the alpha2C adrenergic receptor (ADRA2C) on 4p16. *Human Molecular Genetics*, **1992**, 1, 452	5.6	8
74	Predictive testing for Huntington disease. *Journal of Medical Ethics*, **1992**, 18, 47-8	2.5	
73	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
72	New insights into the clinical features, pathogenesis and molecular genetics of Huntington disease. *Brain Pathology*, **1992**, 2, 321-35	6	45
71	A missense mutation (Asp250----Asn) in exon 6 of the human lipoprotein lipase gene causes chylomicronemia in patients of different ancestries. *Genomics*, **1992**, 13, 649-53	4.3	38
70	Nonrandom association between Huntington disease and two loci separated by about 3 Mb on 4p16.3. *Genomics*, **1992**, 13, 301-11	4.3	29
69	The human beta-subunit of rod photoreceptor cGMP phosphodiesterase: complete retinal cDNA sequence and evidence for expression in brain. *Genomics*, **1992**, 13, 698-704	4.3	32
68	Mapping of the epitope on lipoprotein lipase recognized by a monoclonal antibody (5D2) which inhibits lipase activity. *Lipids and Lipid Metabolism*, **1992**, 1128, 113-5		47
67	Geographic distribution and genealogy of mutation 207 of the lipoprotein lipase gene in the French Canadian population of QuÞbec. *Human Genetics*, **1992**, 89, 671-5	6.3	30
66	Auxiliary-directed dioxygenation: stereoselective synthesis of a diene hydroperoxide. *Tetrahedron Letters*, **1992**, 33, 443-446	2	13
65	Exclusion of DNA changes in the beta-subunit of the c-GMP phosphodiesterase gene as the cause for Huntington's disease. *Nature Genetics*, **1992**, 1, 104-8	36.3	12
64	Delineation of a 50 kilobase DNA segment containing the recombination site in a sporadic case of Huntington's disease. *Nature Genetics*, **1992**, 2, 216-22	36.3	7

63	A missense mutation Pro157 Arg in lipoprotein lipase (LPLNijmegen) resulting in loss of catalytic activity. *FEBS Journal*, **1992**, 208, 267-72		17
62	Linkage studies and mutation analysis of the PDEB gene in 23 families with Leber congenital amaurosis. *Human Mutation*, **1992**, 1, 478-85	4.7	6
61	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
60	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
59	Cortical glucose metabolism in Huntington's disease. *Neurology*, **1992**, 42, 223-9	6.5	67
58	Genetic variants affecting human lipoprotein and hepatic lipases. *Current Opinion in Lipidology*, **1991**, 2, 104-109	4.4	48
57	The FDG/PET methodology for early detection of disease onset: a statistical model. *Journal of Cerebral Blood Flow and Metabolism*, **1991**, 11, A96-102	7.3	35
56	Intrachromosomal location of the telomeric repeat (TTAGGG)n. *Mammalian Genome*, **1991**, 1, 211-6	3.2	27
55	Genomic organization and complete sequence of the human gene encoding the beta-subunit of the cGMP phosphodiesterase and its localisation to 4p 16.3. *Nucleic Acids Research*, **1991**, 19, 6263-8	20.1	56
54	A mutation in the human lipoprotein lipase gene as the most common cause of familial chylomicronemia in French Canadians. *New England Journal of Medicine*, **1991**, 324, 1761-6	59.2	95
53	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
52	Identification of multiple CpG islands and associated conserved sequences in a candidate region for the Huntington disease gene. *Genomics*, **1991**, 11, 1113-24	4.3	19
51	A low-copy repeat located in subtelomeric regions of 14 different human chromosomal termini. *Cytogenetic and Genome Research*, **1991**, 57, 179-83	1.9	10
50	Amino acid substitution (Ile194----Thr) in exon 5 of the lipoprotein lipase gene causes lipoprotein lipase deficiency in three unrelated probands. Support for a multicentric origin. *Journal of Clinical Investigation*, **1991**, 87, 2005-11	15.9	65
49	Regional cerebral glucose metabolism in Turner syndrome. *Canadian Journal of Neurological Sciences*, **1990**, 17, 140-4	1	72
48	DNA testing for Huntington disease results in a modification of risk and not diagnosis of disease. *European Journal of Pediatrics*, **1990**, 149, 513	4.1	1
47	Characterization and organization of DNA sequences adjacent to the human telomere associated repeat (TTAGGG)n. *Nucleic Acids Research*, **1990**, 18, 3353-61	20.1	52
46	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

45	Perinatal and first year follow-up of patients with Prader-Willi syndrome: normal size of hands and feet. *Clinical Genetics*, **1989**, 35, 161-6	4	11
44	Different options for prenatal testing for Huntington's disease using DNA probes. *Journal of Medical Genetics*, **1989**, 26, 353-7	5.8	19
43	Non-random association between alleles detected at D4S95 and D4S98 and the Huntington's disease gene. *Journal of Medical Genetics*, **1989**, 26, 676-81	5.8	67
42	Predictive testing for Huntington disease: I. Description of a pilot project in British Columbia. *American Journal of Medical Genetics Part A*, **1989**, 32, 211-6		79
41	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
40	Epiphyseal dysplasia, microcephaly, nystagmus, and retinitis pigmentosa. *American Journal of Medical Genetics Part A*, **1989**, 33, 341-5		13
39	Huntington disease: no evidence for locus heterogeneity. *Genomics*, **1989**, 5, 304-8	4.3	51
38	Silent periods, long-latency reflexes and cortical MEPs in Huntington's disease and at-risk relatives. *Electroencephalography and Clinical Neurophysiology - Evoked Potentials*, **1989**, 74, 444-9		46
37	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
36	Sequence and expression of Tangier apoA-I gene. *FEBS Journal*, **1988**, 173, 465-71		29
35	A highly polymorphic locus very tightly linked to the Huntington's disease gene. *Nature*, **1988**, 332, 734-6	50.4	155
34	Comparison of gemfibrozil and clofibrate on serum lipids in familial combined hyperlipidemia. A randomized placebo-controlled, double-blind, crossover clinical trial. *Atherosclerosis*, **1988**, 73, 233-40	3.1	9
33	Prenatal diagnosis of asplenia/polysplenia syndrome. *American Journal of Obstetrics and Gynecology*, **1988**, 158, 1085-7	6.4	22
32	Presymptomatic neuropsychological impairment in Huntington's disease. *Archives of Neurology*, **1988**, 45, 769-73		79
31	Hypoalphalipoproteinemia resembling fish eye disease. *Acta Medica Scandinavica*, **1987**, 221, 291-8		22
30	A polymorphic DNA probe located to human chromosome 4p16 (D4S62). *Nucleic Acids Research*, **1987**, 15, 3938	20.1	5
29	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
28	Insufficient evidence to invoke defects in or around the A-I gene as the cause for familial hypoalphalipoproteinemia. *Atherosclerosis*, **1987**, 67, 271-2	3.1	

27	A somatic cell hybrid panel for localizing DNA segments near the Huntington's disease gene. *Genomics*, **1987**, 1, 29-34	4.3	69
26	Cerebral glucose and dopa metabolism in movement disorders. *Canadian Journal of Neurological Sciences*, **1987**, 14, 448-51	1	13
25	Preclinical testing in Huntington disease. *American Journal of Medical Genetics Part A*, **1987**, 27, 733-4		11
24	Ethical issues in preclinical testing in Huntington disease: response to Margery Shaw's invited editorial comment. *American Journal of Medical Genetics Part A*, **1987**, 28, 761-3		4
23	Controlling for cerebral atrophy in positron emission tomography data. *Journal of Cerebral Blood Flow and Metabolism*, **1987**, 7, 510-2	7.3	11
22	Arrested epidermal morphogenesis in three newborn infants with a fatal genetic disorder (restrictive dermopathy). *Journal of Investigative Dermatology*, **1987**, 88, 330-9	4.3	43
21	Restrictive dermopathy: a newly recognized autosomal recessive skin dysplasia. *American Journal of Medical Genetics Part A*, **1986**, 24, 631-48		86
20	Cerebral metabolism of glucose in benign hereditary chorea. *Movement Disorders*, **1986**, 1, 33-44	7	36
19	Regression model for predicting dissociations of regional cerebral glucose metabolism in individuals at risk for Huntington's disease. *Journal of Cerebral Blood Flow and Metabolism*, **1986**, 6, 756-62	7.3	17
18	Positron emission tomography in the early diagnosis of Huntington's disease. *Neurology*, **1986**, 36, 888-94	4.5	142
17	Primary lipoprotein lipase deficiency. *Advances in Experimental Medicine and Biology*, **1986**, 201, 227-39	3.6	24
16	Deletion of Huntington's disease-linked G8 (D4S10) locus in Wolf-Hirschhorn syndrome. *Nature*, **1985**, 318, 75-8	50.4	106
15	Urinary proteins in a patient with Tangier disease. *Clinical Biochemistry*, **1985**, 18, 98-101	3.5	3
14	Bilateral renal agenesis in twins. *American Journal of Medical Genetics Part A*, **1985**, 21, 147-52, 167-9		5
13	Down's syndrome and Alzheimer's disease. *Annals of Neurology*, **1984**, 16, 263	9.4	1
12	Reflections on the history of Huntington's chorea. *Trends in Neurosciences*, **1983**, 6, 122-124	13.3	7
11	The high frequency of juvenile Huntington's chorea in South Africa. *Journal of Medical Genetics*, **1982**, 19, 94-7	5.8	19
10	Genetic aspects of Huntington's chorea: results of a national survey. *American Journal of Medical Genetics Part A*, **1982**, 11, 135-41		17

9	Huntington Chorea **1981**,	228
8	Clinical Features **1981**, 59-92	5
7	Living with Huntington Chorea: The Social Perspective **1981**, 123-135	2
6	Coupled Control of Distal Axon Integrity and Somal Responses to Axonal Damage by the Palmitoyl Acyltransferase ZDHHC17	1
5	Gene expression profiles complement the analysis of genomic modifiers of the clinical onset of Huntington disease	1
4	Feeding schedule and proteolysis regulate autophagic clearance of mutant huntingtin	1
3	Axonal ER Ca2+ Release Enhances Miniature, but Reduces Activity-Dependent Glutamate Release in a Huntington Disease Model	2
2	Length of uninterrupted CAG repeats, independent of polyglutamine size, results in increased somatic instability and hastened age of onset in Huntington disease	4
1	The interaction of aging and oxidative stress contributes to pathogenesis in mouse and human Huntington disease neurons	2