# Harry T Orr

List of Publications by Year
in descending order

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| 169 | 22,192 | 13099 68 | 8866 145 |
|---|---|---|---|
| papers | citations | h-index | g-index |

| 179 | 179 | 179 | 12393 |
|---|---|---|---|
| all docs | docs citations | times ranked | citing authors |

| # | Article | IF | Citations |
|---|---|---|---|
| 1 | Expansion of an unstable trinucleotide CAG repeat in spinocerebellar ataxia type 1. Nature Genetics, 1993, 4, 221-226. | 21.4 | 1,673 |
| 2 | Trinucleotide Repeat Disorders. Annual Review of Neuroscience, 2007, 30, 575-621. | 10.7 | 1,289 |
| 3 | Glutamine Repeats and Neurodegeneration. Annual Review of Neuroscience, 2000, 23, 217-247. | 10.7 | 1,243 |
| 4 | Ataxin-1 Nuclear Localization and Aggregation. Cell, 1998, 95, 41-53. | 28.9 | 965 |
| 5 | Chaperone suppression of aggregation and altered subcellular proteasome localization imply protein misfolding in SCA1. Nature Genetics, 1998, 19, 148-154. | 21.4 | 802 |
| 6 | Identification of genes that modify ataxin-1-induced neurodegeneration. Nature, 2000, 408, 101-106. | 27.8 | 648 |
| 7 | RNAi suppresses polyglutamine-induced neurodegeneration in a model of spinocerebellar ataxia. Nature Medicine, 2004, 10, 816-820. | 30.7 | 643 |
| 8 | SCA1 transgenic mice: A model for neurodegeneration caused by an expanded CAG trinucleotide repeat. Cell, 1995, 82, 937-948. | 28.9 | 567 |
| 9 | Ataxin-1 with an expanded glutamine tract alters nuclear matrix-associated structures. Nature, 1997, 389, 971-974. | 27.8 | 531 |
| 10 | Major histocompatibility antigens: The human (HLA-A,-B,-C) and murine (H-2K, H-2D) class I molecules. Cell, 1981, 24, 287-299. | 28.9 | 517 |
| 11 | Evidence for a mechanism predisposing to intergenerational CAG repeat instability in spinocerebellar ataxia type I. Nature Genetics, 1993, 5, 254-258. | 21.4 | 489 |
| 12 | Mutation of the E6-AP Ubiquitin Ligase Reduces Nuclear Inclusion Frequency While Accelerating Polyglutamine-Induced Pathology in SCA1 Mice. Neuron, 1999, 24, 879-892. | 8.1 | 482 |
| 13 | Interaction of Akt-Phosphorylated Ataxin-1 with 14-3-3 Mediates Neurodegeneration in Spinocerebellar Ataxia Type 1. Cell, 2003, 113, 457-468. | 28.9 | 402 |
| 14 | Identification and characterization of the gene causing type 1 spinocerebellar ataxia. Nature Genetics, 1994, 7, 513-520. | 21.4 | 362 |
| 15 | Polyglutamine expansion down-regulates specific neuronal genes before pathologic changes in SCA1. Nature Neuroscience, 2000, 3, 157-163. | 14.8 | 341 |
| 16 | Spinocerebellar ataxia type 5 in a family descended from the grandparents of President Lincoln maps to chromosome 11. Nature Genetics, 1994, 8, 280-284. | 21.4 | 334 |
| 17 | A Long CAG Repeat in the Mouse Sca1 Locus Replicates SCA1 Features and Reveals the Impact of Protein Solubility on Selective Neurodegeneration. Neuron, 2002, 34, 905-919. | 8.1 | 320 |
| 18 | Serine 776 of Ataxin-1 Is Critical for Polyglutamine-Induced Disease in SCA1 Transgenic Mice. Neuron, 2003, 38, 375-387. | 8.1 | 303 |

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|---|---|---|---|
| 19 | Expression analysis of the ataxinâ€“1 protein in tissues from normal and spinocerebellar ataxia type 1 individuals. Nature Genetics, 1995, 10, 94-98. | 21.4 | 291 |
| 20 | Opposing effects of polyglutamine expansion on native protein complexes contribute to SCA1. Nature, 2008, 452, 713-718. | 27.8 | 287 |
| 21 | Assembly and maturation of HLA-A and HLA-B antigens in vivo. Cell, 1979, 18, 979-991. | 28.9 | 286 |
| 22 | ATAXIN-1 Interacts with the Repressor Capicua in Its Native Complex to Cause SCA1 Neuropathology. Cell, 2006, 127, 1335-1347. | 28.9 | 284 |
| 23 | Polyglutamine spinocerebellar ataxias â€” from genes to potential treatments. Nature Reviews Neuroscience, 2017, 18, 613-626. | 10.2 | 270 |
| 24 | Purkinje Cell Expression of a Mutant Allele ofSCA1in Transgenic Mice Leads to Disparate Effects on Motor Behaviors, Followed by a Progressive Cerebellar Dysfunction and Histological Alterations. Journal of Neuroscience, 1997, 17, 7385-7395. | 3.6 | 261 |
| 25 | Recovery from Polyglutamine-Induced Neurodegeneration in Conditional SCA1 Transgenic Mice. Journal of Neuroscience, 2004, 24, 8853-8861. | 3.6 | 257 |
| 26 | The cerebellar leucine-rich acidic nuclear protein interacts with ataxin-1. Nature, 1997, 389, 974-978. | 27.8 | 246 |
| 27 | Differential expression of HLA-E, HLA-F, and HLA-G transcripts in human tissue. Human Immunology, 1990, 29, 131-142. | 2.4 | 219 |
| 28 | RORÎ±-Mediated Purkinje Cell Development Determines Disease Severity in Adult SCA1 Mice. Cell, 2006, 127, 697-708. | 28.9 | 210 |
| 29 | Pathogenic Mechanisms of a Polyglutamine-mediated Neurodegenerative Disease, Spinocerebellar Ataxia Type 1. Journal of Biological Chemistry, 2009, 284, 7425-7429. | 3.4 | 206 |
| 30 | Disrupted cerebellar cortical development and progressive degeneration of Purkinje cells in SV40 T antigen transgenic mice. Neuron, 1992, 9, 955-966. | 8.1 | 201 |
| 31 | Mice Lacking Ataxin-1 Display Learning Deficits and Decreased Hippocampal Paired-Pulse Facilitation. Journal of Neuroscience, 1998, 18, 5508-5516. | 3.6 | 197 |
| 32 | miR-19, miR-101 and miR-130 co-regulate ATXN1 levels to potentially modulate SCA1 pathogenesis. Nature Neuroscience, 2008, 11, 1137-1139. | 14.8 | 194 |
| 33 | The Unstable Repeatsâ€”Three Evolving Faces of Neurological Disease. Neuron, 2013, 77, 825-843. | 8.1 | 192 |
| 34 | The AXH Domain of Ataxin-1 Mediates Neurodegeneration through Its Interaction with Gfi-1/Senseless Proteins. Cell, 2005, 122, 633-644. | 28.9 | 189 |
| 35 | The GSK3Î² signaling cascade and neurodegenerative disease. Current Opinion in Neurobiology, 2002, 12, 275-278. | 4.2 | 188 |
| 36 | Gametic and somatic tissueâ€“specific heterogeneity of the expanded SCA1 CAG repeat in spinocerebellar ataxia type 1. Nature Genetics, 1995, 10, 344-350. | 21.4 | 179 |

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| 37 | Tolerance is established in polyclonal CD4+ T cells by distinct mechanisms, according to self-peptide expression patterns. Nature Immunology, 2016, 17, 187-195. | 14.5 | 178 |
| 38 | Gene profiling links SCA1 pathophysiology to glutamate signaling in Purkinje cells of transgenic mice. Human Molecular Genetics, 2004, 13, 2535-2543. | 2.9 | 168 |
| 39 | HLA and maternalâ€fetal recognition. FASEB Journal, 1992, 6, 2344-2348. | 0.5 | 151 |
| 40 | Lithium Therapy Improves Neurological Function and Hippocampal Dendritic Arborization in a Spinocerebellar Ataxia Type 1 Mouse Model. PLoS Medicine, 2007, 4, e182. | 8.4 | 147 |
| 41 | Exercise and Genetic Rescue of SCA1 via the Transcriptional Repressor Capicua. Science, 2011, 334, 690-693. | 12.6 | 144 |
| 42 | Cell biology of spinocerebellar ataxia. Journal of Cell Biology, 2012, 197, 167-177. | 5.2 | 144 |
| 43 | Pumilio1 Haploinsufficiency Leads to SCA1-like Neurodegeneration by Increasing Wild-Type Ataxin1 Levels. Cell, 2015, 160, 1087-1098. | 28.9 | 139 |
| 44 | SCA1-like Disease in Mice Expressing Wild-Type Ataxin-1 with a Serine to Aspartic Acid Replacement at Residue 776. Neuron, 2010, 67, 929-935. | 8.1 | 137 |
| 45 | Aminopyridines Correct Early Dysfunction and Delay Neurodegeneration in a Mouse Model of Spinocerebellar Ataxia Type 1. Journal of Neuroscience, 2011, 31, 11795-11807. | 3.6 | 137 |
| 46 | The heavy chain of human histocompatibility antigen HLA-B7 contains an immunoglobulin-like region. Nature, 1979, 282, 266-270. | 27.8 | 134 |
| 47 | Polyglutamine neurodegenerative diseases and regulation of transcription: assembling the puzzle. Genes and Development, 2006, 20, 2183-2192. | 5.9 | 129 |
| 48 | Complete amino acid sequence of a papain-solubilized human histocompatibility antigen, HLA-B7. 2. Sequence determination and search for homologies. Biochemistry, 1979, 18, 5711-5720. | 2.5 | 127 |
| 49 | Spinocerebellar ataxia type 1. Seminars in Cell Biology, 1995, 6, 29-35. | 3.4 | 125 |
| 50 | RASâ€“MAPKâ€“MSK1 pathway modulates ataxin 1 protein levels and toxicity in SCA1. Nature, 2013, 498, 325-331. | 27.8 | 119 |
| 51 | Partial Loss of Ataxin-1 Function Contributes to Transcriptional Dysregulation in Spinocerebellar Ataxia Type 1 Pathogenesis. PLoS Genetics, 2010, 6, e1001021. | 3.5 | 113 |
| 52 | Disruption of the ATXN1â€“CIC complex causes a spectrum of neurobehavioral phenotypes in mice and humans. Nature Genetics, 2017, 49, 527-536. | 21.4 | 113 |
| 53 | RNA association and nucleocytoplasmic shuttling by ataxin-1. Journal of Cell Science, 2005, 118, 233-242. | 2.0 | 109 |
| 54 | Antisense oligonucleotideâ€“mediated ataxin-1 reduction prolongs survival in SCA1 mice and reveals disease-associated transcriptome profiles. JCI Insight, 2018, 3, . | 5.0 | 106 |

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| 55 | Polyglutamine diseases: protein cleavage and aggregation. Current Opinion in Neurobiology, 1999, 9, 566-570. | 4.2 | 102 |
| 56 | Increased Trinucleotide Repeat Instability with Advanced Maternal Age. Human Molecular Genetics, 1997, 6, 2135-2139. | 2.9 | 97 |
| 57 | Neuronal Atrophy Early in Degenerative Ataxia Is a Compensatory Mechanism to Regulate Membrane Excitability. Journal of Neuroscience, 2015, 35, 11292-11307. | 3.6 | 93 |
| 58 | ATXN1 Protein Family and CIC Regulate Extracellular Matrix Remodeling and Lung Alveolarization. Developmental Cell, 2011, 21, 746-757. | 7.0 | 89 |
| 59 | SUMOylation of the Polyglutamine Repeat Protein, Ataxin-1, Is Dependent on a Functional Nuclear Localization Signal. Journal of Biological Chemistry, 2005, 280, 21942-21948. | 3.4 | 87 |
| 60 | Cerebellar Transcriptome Profiles of ATXN1 Transgenic Mice Reveal SCA1 Disease Progression and Protection Pathways. Neuron, 2016, 89, 1194-1207. | 8.1 | 86 |
| 61 | The insulin-like growth factor pathway is altered in spinocerebellar ataxia type 1 and type 7. Proceedings of the National Academy of Sciences of the United States of America, 2008, 105, 1291-1296. | 7.1 | 85 |
| 62 | Noninvasive Detection of Presymptomatic and Progressive Neurodegeneration in a Mouse Model of Spinocerebellar Ataxia Type 1. Journal of Neuroscience, 2010, 30, 3831-3838. | 3.6 | 85 |
| 63 | Use of HLA loss mutants to analyse the structure of the human major histocompatibility complex. Nature, 1982, 296, 454-456. | 27.8 | 84 |
| 64 | Altered Trafficking of Membrane Proteins in Purkinje Cells of SCA1 Transgenic Mice. American Journal of Pathology, 2001, 159, 905-913. | 3.8 | 83 |
| 65 | Reversing Neurodegeneration:A Promise Unfolds. Cell, 2000, 101, 1-4. | 28.9 | 81 |
| 66 | ATXN1-CIC Complex Is the Primary Driver of Cerebellar Pathology in Spinocerebellar Ataxia Type 1 through a Gain-of-Function Mechanism. Neuron, 2018, 97, 1235-1243.e5. | 8.1 | 79 |
| 67 | Emerging pathogenic pathways in the spinocerebellar ataxias. Current Opinion in Genetics and Development, 2009, 19, 247-253. | 3.3 | 77 |
| 68 | Duplication of Atxn1l suppresses SCA1 neuropathology by decreasing incorporation of polyglutamine-expanded ataxin-1 into native complexes. Nature Genetics, 2007, 39, 373-379. | 21.4 | 75 |
| 69 | Abnormalities in the Climbing Fiber-Purkinje Cell Circuitry Contribute to Neuronal Dysfunction inATXN1[82Q] Mice. Journal of Neuroscience, 2011, 31, 12778-12789. | 3.6 | 75 |
| 70 | In Vivo Viability of Postmitotic Purkinje Neurons Requires pRb Family Member Function. Molecular and Cellular Neurosciences, 1995, 6, 153-167. | 2.2 | 70 |
| 71 | Targeted Deletion of a Single Sca8 Ataxia Locus Allele in Mice Causes Abnormal Gait, Progressive Loss of Motor Coordination, and Purkinje Cell Dendritic Deficits. Journal of Neuroscience, 2006, 26, 9975-9982. | 3.6 | 70 |
| 72 | Polyglutamine neurodegeneration: expanded glutamines enhance native functions. Current Opinion in Genetics and Development, 2012, 22, 251-255. | 3.3 | 69 |

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| 73 | Class I-like HLA genes map telomeric to the HLA-A2 locus in human cells. Nature, 1983, 302, 534-536. | 27.8 | 68 |
| 74 | Extensive cryptic splicing upon loss of RBM17 and TDP43 in neurodegeneration models. Human Molecular Genetics, 2016, 25, ddw337. | 2.9 | 68 |
| 75 | Progress in pathogenesis studies of spinocerebellar ataxia type 1. Philosophical Transactions of the Royal Society B: Biological Sciences, 1999, 354, 1079-1081. | 4.0 | 66 |
| 76 | Regional rescue of spinocerebellar ataxia type 1 phenotypes by 14-3-3 Îµ haploinsufficiency in mice underscores complex pathogenicity in neurodegeneration. Proceedings of the National Academy of Sciences of the United States of America, 2011, 108, 2142-2147. | 7.1 | 65 |
| 77 | HLA non-A,B,C class I genes: Their structure and expression. Immunologic Research, 1990, 9, 265-274. | 2.9 | 63 |
| 78 | Mapmodulin/Leucine-rich Acidic Nuclear Protein Binds the Light Chain of Microtubule-associated Protein 1B and Modulates Neuritogenesis. Journal of Biological Chemistry, 2003, 278, 34691-34699. | 3.4 | 62 |
| 79 | Cloning and Developmental Expression Analysis of the Murine Homolog of the Spinocerebellar Ataxia Type 1 Gene (Sca1). Human Molecular Genetics, 1996, 5, 33-40. | 2.9 | 59 |
| 80 | Organization of the human class I major histocompatibility complex genes. Immunologic Research, 1987, 6, 1-10. | 2.9 | 58 |
| 81 | Partial loss of Tip60 slows mid-stage neurodegeneration in a spinocerebellar ataxia type 1 (SCA1) mouse model. Human Molecular Genetics, 2011, 20, 2204-2212. | 2.9 | 58 |
| 82 | Calcium Dynamics and Electrophysiological Properties of Cerebellar Purkinje Cells in SCA1 Transgenic Mice. Journal of Neurophysiology, 2001, 85, 1750-1760. | 1.8 | 57 |
| 83 | Phosphorylation of ATXN1 at Ser776 in the cerebellum. Journal of Neurochemistry, 2009, 110, 675-686. | 3.9 | 55 |
| 84 | Regional differences of somatic CAG repeat instability do not account for selective neuronal vulnerability in a knock-in mouse model of SCA1. Human Molecular Genetics, 2003, 12, 2789-2795. | 2.9 | 54 |
| 85 | Molecular analysis of the variant alloantigen HLA-B27d (HLA-Bâ^—2703) identifies a unique single amino acid substitution. Human Immunology, 1988, 21, 209-219. | 2.4 | 53 |
| 86 | Purkinje Cell Ataxin-1 Modulates Climbing Fiber Synaptic Input in Developing and Adult Mouse Cerebellum. Journal of Neuroscience, 2013, 33, 5806-5820. | 3.6 | 50 |
| 87 | Mapping of class I DNA sequences within the human major histocompatibility complex. Immunogenetics, 1983, 18, 489-502. | 2.4 | 49 |
| 88 | Spinocerebellar Ataxia Type 1â€"Modeling the Pathogenesis of a Polyglutamine Neurodegenerative Disorder in Transgenic Mice. Journal of Neuropathology and Experimental Neurology, 2000, 59, 265-270. | 1.7 | 49 |
| 89 | 14-3-3 Binding to Ataxin-1(ATXN1) Regulates Its Dephosphorylation at Ser-776 and Transport to the Nucleus. Journal of Biological Chemistry, 2011, 286, 34606-34616. | 3.4 | 49 |
| 90 | The Effects of the Polyglutamine Repeat Protein Ataxin-1 on the UbL-UBA Protein A1Up. Journal of Biological Chemistry, 2004, 279, 42290-42301. | 3.4 | 44 |

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| 91 | Reduction of Purkinje Cell Pathology in SCA1 Transgenic Mice by p53 Deletion. Neurobiology of Disease, 2001, 8, 974-981. | 4.4 | 39 |
| 92 | Generation and Characterization of LANP/pp32 Null Mice. Molecular and Cellular Biology, 2004, 24, 3140-3149. | 2.3 | 38 |
| 93 | Complete amino acid sequence of a papain-solubilized human histocompatibility antigen, HLA-B7. 1. Isolation and amino acid composition of fragments and of tryptic and chymotryptic peptides. Biochemistry, 1979, 18, 5704-5711. | 2.5 | 36 |
| 94 | SCA1â€"Phosphorylation, a regulator of Ataxin-1 function and pathogenesis. Progress in Neurobiology, 2012, 99, 179-185. | 5.7 | 33 |
| 95 | RNA Targets of the Fragile X Protein. Cell, 2001, 107, 555-557. | 28.9 | 30 |
| 96 | Neuron protection agency. Nature, 2004, 431, 747-748. | 27.8 | 30 |
| 97 | Polyglutamine Disease Toxicity Is Regulated by Nemo-like Kinase in Spinocerebellar Ataxia Type 1. Journal of Neuroscience, 2013, 33, 9328-9336. | 3.6 | 30 |
| 98 | HLA-G Transgenic Mice: A Model for Studying Expression and Function at the Maternal/Fetal Interface. Immunological Reviews, 1995, 147, 53-65. | 6.0 | 29 |
| 99 | In vivo monitoring of recovery from neurodegeneration in conditional transgenic SCA1 mice. Experimental Neurology, 2011, 232, 290-298. | 4.1 | 29 |
| 100 | A native interactor scaffolds and stabilizes toxic ATAXIN-1 oligomers in SCA1. ELife, 2015, 4, . | 6.0 | 29 |
| 101 | FTD and ALS: Genetic Ties that Bind. Neuron, 2011, 72, 189-190. | 8.1 | 28 |
| 102 | The Transcription Factor E2F-1 in SV40 T Antigen-Induced Cerebellar Purkinje Cell Degeneration. Molecular and Cellular Neurosciences, 1998, 12, 16-28. | 2.2 | 27 |
| 103 | A cell-based screen for modulators of ataxin-1 phosphorylation. Human Molecular Genetics, 2005, 14, 1095-1105. | 2.9 | 27 |
| 104 | Reduction of protein kinase A-mediated phosphorylation of ATXN1-S776 in Purkinje cells delays onset of Ataxia in a SCA1 mouse model. Neurobiology of Disease, 2018, 116, 93-105. | 4.4 | 27 |
| 105 | Targeting inhibitory cerebellar circuitry to alleviate behavioral deficits in a mouse model for studying idiopathic autism. Neuropsychopharmacology, 2020, 45, 1159-1170. | 5.4 | 26 |
| 106 | Assessing recovery from neurodegeneration in spinocerebellar ataxia 1: Comparison of in vivo magnetic resonance spectroscopy with motor testing, gene expression and histology. Neurobiology of Disease, 2015, 74, 158-166. | 4.4 | 25 |
| 107 | Susceptibility to Cell Death Induced by Mutant SV40 T-Antigen Correlates with Purkinje Neuron Functional Development. Molecular and Cellular Neurosciences, 1997, 9, 42-62. | 2.2 | 24 |
| 108 | Motor neuron degeneration correlates with respiratory dysfunction in SCA1. DMM Disease Models and Mechanisms, 2018, 11, . | 2.4 | 23 |

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| 109 | The Ins and Outs of a Polyglutamine Neurodegenerative Disease: Spinocerebellar Ataxia Type 1 (SCA1). Neurobiology of Disease, 2000, 7, 129-134. | 4.4 | 22 |
| 110 | Amino Acids in a Region of Ataxin-1 Outside of the Polyglutamine Tract Influence the Course of Disease in SCA1 Transgenic Mice. NeuroMolecular Medicine, 2002, 1, 33-42. | 3.4 | 22 |
| 111 | Diminishing return for mechanistic therapeutics with neurodegenerative disease duration?. BioEssays, 2016, 38, 977-980. | 2.5 | 22 |
| 112 | Linkage of an Alzheimer disease susceptibility locus to markers on human chromosome 21. American Journal of Medical Genetics Part A, 1991, 40, 449-453. | 2.4 | 21 |
| 113 | Protocol for genetic testing in Huntington disease: Three years of experience in Minnesota. American Journal of Medical Genetics Part A, 1991, 40, 518-522. | 2.4 | 21 |
| 114 | Genetically engineered mouse models of the trinucleotide-repeat spinocerebellar ataxias. Brain Research Bulletin, 2012, 88, 33-42. | 3.0 | 21 |
| 115 | Cytotoxic T Lymphocyte Recognition of HLA-G in Mice. Human Immunology, 1997, 55, 127-139. | 2.4 | 20 |
| 116 | Mouse Models of Human CAG Repeat Disorders. Brain Pathology, 1997, 7, 965-977. | 4.1 | 20 |
| 117 | Altered Capicua expression drives regional Purkinje neuron vulnerability through ion channel gene dysregulation in spinocerebellar ataxia type 1. Human Molecular Genetics, 2020, 29, 3249-3265. | 2.9 | 20 |
| 118 | Molecular and endocrine characterization of a mutation involving a recombination between the steroid 21-hydroxylase functional gene and pseudogene. Journal of Steroid Biochemistry and Molecular Biology, 1991, 38, 677-686. | 2.5 | 19 |
| 119 | Spinocerebellar Ataxia Type 1: Molecular Mechanisms of Neurodegeneration and Preclinical Studies. Advances in Experimental Medicine and Biology, 2018, 1049, 135-145. | 1.6 | 19 |
| 120 | Characterization of the Zebrafishatxn1/axhGene Family. Journal of Neurogenetics, 2009, 23, 313-323. | 1.4 | 18 |
| 121 | Use of DNA probes from the 5â€² flanking region of the HLA-B gene to examine polymorphism at the HLA-B locus. Human Immunology, 1986, 16, 137-147. | 2.4 | 16 |
| 122 | Diagnosis of classical steroid 21-hydroxylase deficiency using an HLA-B locus-specific DNA-probe. American Journal of Medical Genetics Part A, 1988, 29, 703-712. | 2.4 | 16 |
| 123 | PAK1 regulates ATXN1 levels providing an opportunity to modify its toxicity in spinocerebellar ataxia type 1. Human Molecular Genetics, 2018, 27, 2863-2873. | 2.9 | 16 |
| 124 | Antisense Oligonucleotide Therapeutic Approach for Suppression of Ataxin-1 Expression: A Safety Assessment. Molecular Therapy - Nucleic Acids, 2020, 21, 1006-1016. | 5.1 | 16 |
| 125 | Ataxin-1 oligomers induce local spread of pathology and decreasing them by passive immunization slows Spinocerebellar ataxia type 1 phenotypes. ELife, 2015, 4, . | 6.0 | 16 |
| 126 | Autistic-like behavior and cerebellar dysfunction in Bmal1 mutant mice ameliorated by mTORC1 inhibition. Molecular Psychiatry, 2023, 28, 3727-3738. | 7.9 | 16 |

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| 127 | Microarrays and polyglutamine disorders: reports from the Hereditary Disease Array Group. Human Molecular Genetics, 2002, 11, 1909-1910. | 2.9 | 15 |
| 128 | Identification of a novel phosphorylation site in ataxin-1. Biochimica Et Biophysica Acta - Molecular Cell Research, 2005, 1744, 11-18. | 4.1 | 15 |
| 129 | Visualizing and mapping the cerebellum with serial optical coherence scanner. Neurophotonics, 2016, 4, 1. | 3.3 | 15 |
| 130 | The ataxin-1 interactome reveals direct connection with multiple disrupted nuclear transport pathways. Nature Communications, 2020, 11, 3343. | 12.8 | 15 |
| 131 | Consensus Paper: Strengths and Weaknesses of Animal Models of Spinocerebellar Ataxias and Their Clinical Implications. Cerebellum, 2022, 21, 452-481. | 2.5 | 15 |
| 132 | Overexpression of CREB reduces CRE-mediated transcription: behavioral and cellular analyses in transgenic mice. Molecular and Cellular Neurosciences, 2004, 25, 602-611. | 2.2 | 12 |
| 133 | Hsp70/Hsc70 regulates the effect phosphorylation has on stabilizing ataxin-1. Journal of Neurochemistry, 2007, 102, 2040-2048. | 3.9 | 12 |
| 134 | Modulation of ATXN1 S776 phosphorylation reveals the importance of allele-specific targeting in SCA1. JCI Insight, 2021, 6, . | 5.0 | 12 |
| 135 | Unstable trinucleotide repeats and the diagnosis of neurodegenerative disease. Human Pathology, 1994, 25, 598-601. | 2.0 | 11 |
| 136 | Treadmill training increases the motor activity and neuron survival of the cerebellum in a mouse model of spinocerebellar ataxia type 1. Kaohsiung Journal of Medical Sciences, 2019, 35, 679-685. | 1.9 | 11 |
| 137 | Dual targeting of brain regionâ€specific kinases potentiates neurological rescue in Spinocerebellar ataxia type 1. EMBO Journal, 2021, 40, e106106. | 7.8 | 11 |
| 138 | Cholecystokinin 1 receptor activation restores normal mTORC1 signaling and is protective to Purkinje cells of SCA mice. Cell Reports, 2021, 37, 109831. | 6.4 | 11 |
| 139 | Sequence of a murine cDNA, pcp-4, that encodes the homolog of the rat brain-specific antigen PEP-19. Nucleic Acids Research, 1990, 18, 1304-1304. | 14.5 | 10 |
| 140 | Pathogenesis of Polyglutamine-Induced Disease: A Model for SCA1. Molecular Genetics and Metabolism, 1999, 66, 172-178. | 1.1 | 10 |
| 141 | Polarization-sensitive optical coherence tomography reveals gray matter and white matter atrophy in SCA1 mouse models. Neurobiology of Disease, 2018, 116, 69-77. | 4.4 | 10 |
| 142 | Regulation of Ataxin-1 Phosphorylation and Its Impact on Biology. Methods in Molecular Biology, 2013, 1010, 201-209. | 0.9 | 10 |
| 143 | HLA Class I Gene Family: Characterization of Genes Encoding Non-HLA-A,B,C Proteins. , 1989, , 33-40. | | 10 |
| 144 | Qs in the Nucleus. Neuron, 2001, 31, 875-876. | 8.1 | 9 |

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| 145 | RNA gains a new function: a mediator of neurodegeneration. Trends in Neurosciences, 2004, 27, 233-234. | 8.6 | 9 |
| 146 | Lurcher, nPIST, and Autophagy. Neuron, 2002, 35, 813-814. | 8.1 | 8 |
| 147 | Nuclear Ataxias. Cold Spring Harbor Perspectives in Biology, 2010, 2, a000786-a000786. | 5.5 | 8 |
| 148 | Toxic RNA as a driver of disease in a common form of ALS and dementia. Proceedings of the National Academy of Sciences of the United States of America, 2013, 110, 7533-7534. | 7.1 | 8 |
| 149 | Antisense RNA Sequences Modulating the Ataxin-1 Message: Molecular Model of Gene Therapy for Spinocerebellar Ataxia Type 1, a Dominant-Acting Unstable Trinucleotide Repeat Disease. Cell Transplantation, 2008, 17, 723-734. | 2.5 | 7 |
| 150 | Unstable Nucleotide Repeat Minireview Series: A Molecular Biography of Unstable Repeat Disorders. Journal of Biological Chemistry, 2009, 284, 7405. | 3.4 | 7 |
| 151 | Reduction of mutant ATXN1 rescues premature death in a conditional SCA1 mouse model. JCI Insight, 2022, 7, . | 5.0 | 6 |
| 152 | Cross-species genetic screens identify transglutaminase 5 as a regulator of polyglutamine-expanded ataxin-1. Journal of Clinical Investigation, 2022, 132, . | 8.2 | 6 |
| 153 | The Major Histocompatibility Complex: Analysis at the Protein and DNA Levels. , 1982, , 1-51. | | 5 |
| 154 | Into the depths of ataxia. Journal of Clinical Investigation, 2004, 113, 505-507. | 8.2 | 5 |
| 155 | Chapter 4 Complete Primary Structure of Human Histocompatibility Antigen Hla-B7: Evolutionary and Functional Implications. Current Topics in Developmental Biology, 1980, 14, 97-113. | 2.2 | 3 |
| 156 | Are Polyglutamine Diseases Expanding?. Neuron, 2011, 70, 377-378. | 8.1 | 3 |
| 157 | Stephen T. Warren, Ph.D. (1953â€“2021): A remembrance. American Journal of Human Genetics, 2022, 109, 3-11. | 6.2 | 2 |
| 158 | Cholecystokinin Activation of Cholecystokinin 1 Receptors: a Purkinje Cell Neuroprotective Pathway. Cerebellum, 2023, 22, 756-760. | 2.5 | 2 |
| 159 | Complete Primary Structure of Human Histocompatibility Antigen HLA-B7. , 1981, , 479-493. | | 1 |
| 160 | Transfer and Expression of Human Non-A,B,C Class I Genes in Human HLA A,B,C Null Lymphoblastoid Cells. , 1989, , 159-161. | | 1 |
| 161 | Animal Models of Spinocerebellar Ataxia Type 1 (SCA1). , 2005, , 623-630. | | 0 |
| 162 | Animal Models of Spinocerebellar Ataxia Type 1. , 2015, , 979-990. | | 0 |

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