Harry T Orr

List of Publications by Citations

167 papers	19,253 citations	65 h-index	138 g-index
179 ext. papers	20,855 ext. citations	15.5 avg, IF	6.49 L-index

#	Paper	IF	Citations
167	Expansion of an unstable trinucleotide CAG repeat in spinocerebellar ataxia type 1. *Nature Genetics*, **1993**, 4, 221-6	36.3	1453
166	Trinucleotide repeat disorders. *Annual Review of Neuroscience*, **2007**, 30, 575-621	17	1120
165	Glutamine repeats and neurodegeneration. *Annual Review of Neuroscience*, **2000**, 23, 217-47	17	1110
164	Ataxin-1 nuclear localization and aggregation: role in polyglutamine-induced disease in SCA1 transgenic mice. *Cell*, **1998**, 95, 41-53	56.2	898
163	Chaperone suppression of aggregation and altered subcellular proteasome localization imply protein misfolding in SCA1. *Nature Genetics*, **1998**, 19, 148-54	36.3	766
162	RNAi suppresses polyglutamine-induced neurodegeneration in a model of spinocerebellar ataxia. *Nature Medicine*, **2004**, 10, 816-20	50.5	577
161	Identification of genes that modify ataxin-1-induced neurodegeneration. *Nature*, **2000**, 408, 101-6	50.4	560
160	SCA1 transgenic mice: a model for neurodegeneration caused by an expanded CAG trinucleotide repeat. *Cell*, **1995**, 82, 937-48	56.2	505
159	Ataxin-1 with an expanded glutamine tract alters nuclear matrix-associated structures. *Nature*, **1997**, 389, 971-4	50.4	498
158	Major histocompatibility antigens: the human (HLA-A, -B, -C) and murine (H-2K, H-2D) class I molecules. *Cell*, **1981**, 24, 287-99	56.2	483
157	Mutation of the E6-AP ubiquitin ligase reduces nuclear inclusion frequency while accelerating polyglutamine-induced pathology in SCA1 mice. *Neuron*, **1999**, 24, 879-92	13.9	449
156	Evidence for a mechanism predisposing to intergenerational CAG repeat instability in spinocerebellar ataxia type I. *Nature Genetics*, **1993**, 5, 254-8	36.3	436
155	Interaction of Akt-phosphorylated ataxin-1 with 14-3-3 mediates neurodegeneration in spinocerebellar ataxia type 1. *Cell*, **2003**, 113, 457-68	56.2	344
154	Identification and characterization of the gene causing type 1 spinocerebellar ataxia. *Nature Genetics*, **1994**, 7, 513-20	36.3	310
153	Polyglutamine expansion down-regulates specific neuronal genes before pathologic changes in SCA1. *Nature Neuroscience*, **2000**, 3, 157-63	25.5	299
152	Spinocerebellar ataxia type 5 in a family descended from the grandparents of President Lincoln maps to chromosome 11. *Nature Genetics*, **1994**, 8, 280-4	36.3	294
151	Serine 776 of ataxin-1 is critical for polyglutamine-induced disease in SCA1 transgenic mice. *Neuron*, **2003**, 38, 375-87	13.9	275

150	Expression analysis of the ataxin-1 protein in tissues from normal and spinocerebellar ataxia type 1 individuals. *Nature Genetics*, **1995**, 10, 94-8	36.3	265
149	Assembly and maturation of HLA-A and HLA-B antigens in vivo. *Cell*, **1979**, 18, 979-91	56.2	262
148	Opposing effects of polyglutamine expansion on native protein complexes contribute to SCA1. *Nature*, **2008**, 452, 713-8	50.4	250
147	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, 905-19	13.9	250
146	ATAXIN-1 interacts with the repressor Capicua in its native complex to cause SCA1 neuropathology. *Cell*, **2006**, 127, 1335-47	56.2	242
145	Purkinje cell expression of a mutant allele of SCA1 in transgenic mice leads to disparate effects on motor behaviors, followed by a progressive cerebellar dysfunction and histological alterations. *Journal of Neuroscience*, **1997**, 17, 7385-95	6.6	233
144	Recovery from polyglutamine-induced neurodegeneration in conditional SCA1 transgenic mice. *Journal of Neuroscience*, **2004**, 24, 8853-61	6.6	229
143	The cerebellar leucine-rich acidic nuclear protein interacts with ataxin-1. *Nature*, **1997**, 389, 974-8	50.4	227
142	Differential expression of HLA-E, HLA-F, and HLA-G transcripts in human tissue. *Human Immunology* , **1990**, 29, 131-42	2.3	195
141	Disrupted cerebellar cortical development and progressive degeneration of Purkinje cells in SV40 T antigen transgenic mice. *Neuron*, **1992**, 9, 955-66	13.9	191
140	miR-19, miR-101 and miR-130 co-regulate ATXN1 levels to potentially modulate SCA1 pathogenesis. *Nature Neuroscience*, **2008**, 11, 1137-9	25.5	182
139	Mice lacking ataxin-1 display learning deficits and decreased hippocampal paired-pulse facilitation. *Journal of Neuroscience*, **1998**, 18, 5508-16	6.6	181
138	RORalpha-mediated Purkinje cell development determines disease severity in adult SCA1 mice. *Cell* , **2006**, 127, 697-708	56.2	180
137	Polyglutamine spinocerebellar ataxias - from genes to potential treatments. *Nature Reviews Neuroscience*, **2017**, 18, 613-626	13.5	174
136	The GSK3 beta signaling cascade and neurodegenerative disease. *Current Opinion in Neurobiology*, **2002**, 12, 275-8	7.6	168
135	Pathogenic mechanisms of a polyglutamine-mediated neurodegenerative disease, spinocerebellar ataxia type 1. *Journal of Biological Chemistry*, **2009**, 284, 7425-9	5.4	165
134	The AXH domain of Ataxin-1 mediates neurodegeneration through its interaction with Gfi-1/Senseless proteins. *Cell*, **2005**, 122, 633-44	56.2	164
133	Gametic and somatic tissue-specific heterogeneity of the expanded SCA1 CAG repeat in spinocerebellar ataxia type 1. *Nature Genetics*, **1995**, 10, 344-50	36.3	163

132	Gene profiling links SCA1 pathophysiology to glutamate signaling in Purkinje cells of transgenic mice. *Human Molecular Genetics*, **2004**, 13, 2535-43	5.6	152
131	The unstable repeats--three evolving faces of neurological disease. *Neuron*, **2013**, 77, 825-43	13.9	151
130	HLA and maternal-fetal recognition. *FASEB Journal*, **1992**, 6, 2344-8	0.9	139
129	Lithium therapy improves neurological function and hippocampal dendritic arborization in a spinocerebellar ataxia type 1 mouse model. *PLoS Medicine*, **2007**, 4, e182	11.6	129
128	Cell biology of spinocerebellar ataxia. *Journal of Cell Biology*, **2012**, 197, 167-77	7.3	123
127	Polyglutamine neurodegenerative diseases and regulation of transcription: assembling the puzzle. *Genes and Development*, **2006**, 20, 2183-92	12.6	121
126	Complete amino acid sequence of a papain-solubilized human histocompatibility antigen, HLA-B7. 2. Sequence determination and search for homologies. *Biochemistry*, **1979**, 18, 5711-20	3.2	121
125	Tolerance is established in polyclonal CD4(+) T cells by distinct mechanisms, according to self-peptide expression patterns. *Nature Immunology*, **2016**, 17, 187-95	19.1	120
124	SCA1-like disease in mice expressing wild-type ataxin-1 with a serine to aspartic acid replacement at residue 776. *Neuron*, **2010**, 67, 929-35	13.9	120
123	The heavy chain of human histocompatibility antigen HLA-B7 contains an immunoglobulin-like region. *Nature*, **1979**, 282, 266-70	50.4	118
122	Exercise and genetic rescue of SCA1 via the transcriptional repressor Capicua. *Science*, **2011**, 334, 690-3	33.3	117
121	Aminopyridines correct early dysfunction and delay neurodegeneration in a mouse model of spinocerebellar ataxia type 1. *Journal of Neuroscience*, **2011**, 31, 11795-807	6.6	104
120	RAS-MAPK-MSK1 pathway modulates ataxin 1 protein levels and toxicity in SCA1. *Nature*, **2013**, 498, 325-331	50.4	101
119	Pumilio1 haploinsufficiency leads to SCA1-like neurodegeneration by increasing wild-type Ataxin1 levels. *Cell*, **2015**, 160, 1087-98	56.2	100
118	Partial loss of ataxin-1 function contributes to transcriptional dysregulation in spinocerebellar ataxia type 1 pathogenesis. *PLoS Genetics*, **2010**, 6, e1001021	6	95
117	Polyglutamine diseases: protein cleavage and aggregation. *Current Opinion in Neurobiology*, **1999**, 9, 566-70	7.6	94
116	RNA association and nucleocytoplasmic shuttling by ataxin-1. *Journal of Cell Science*, **2005**, 118, 233-42	5.3	92
115	Spinocerebellar ataxia type 1. *Seminars in Cell Biology*, **1995**, 6, 29-35		88

114	Increased trinucleotide repeat instability with advanced maternal age. *Human Molecular Genetics*, **1997**, 6, 2135-9	5.6	87
113	Altered trafficking of membrane proteins in purkinje cells of SCA1 transgenic mice. *American Journal of Pathology*, **2001**, 159, 905-13	5.8	80
112	Use of HLA loss mutants to analyse the structure of the human major histocompatibility complex. *Nature*, **1982**, 296, 454-6	50.4	80
111	SUMOylation of the polyglutamine repeat protein, ataxin-1, is dependent on a functional nuclear localization signal. *Journal of Biological Chemistry*, **2005**, 280, 21942-8	5.4	77
110	Reversing neurodegeneration: a promise unfolds. *Cell*, **2000**, 101, 1-4	56.2	75
109	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, 1291-6	11.5	74
108	ATXN1 protein family and CIC regulate extracellular matrix remodeling and lung alveolarization. *Developmental Cell*, **2011**, 21, 746-57	10.2	73
107	Noninvasive detection of presymptomatic and progressive neurodegeneration in a mouse model of spinocerebellar ataxia type 1. *Journal of Neuroscience*, **2010**, 30, 3831-8	6.6	72
106	Disruption of the ATXN1-CIC complex causes a spectrum of neurobehavioral phenotypes in mice and humans. *Nature Genetics*, **2017**, 49, 527-536	36.3	71
105	Neuronal Atrophy Early in Degenerative Ataxia Is a Compensatory Mechanism to Regulate Membrane Excitability. *Journal of Neuroscience*, **2015**, 35, 11292-307	6.6	71
104	Class I-like HLA genes map telomeric to the HLA-A2 locus in human cells. *Nature*, **1983**, 302, 534-6	50.4	66
103	In vivo viability of postmitotic Purkinje neurons requires pRb family member function. *Molecular and Cellular Neurosciences*, **1995**, 6, 153-67	4.8	65
102	Duplication of Atxn1l suppresses SCA1 neuropathology by decreasing incorporation of polyglutamine-expanded ataxin-1 into native complexes. *Nature Genetics*, **2007**, 39, 373-9	36.3	64
101	Emerging pathogenic pathways in the spinocerebellar ataxias. *Current Opinion in Genetics and Development*, **2009**, 19, 247-53	4.9	62
100	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, 9975-82	6.6	62
99	Polyglutamine neurodegeneration: expanded glutamines enhance native functions. *Current Opinion in Genetics and Development*, **2012**, 22, 251-5	4.9	61
98	Abnormalities in the climbing fiber-Purkinje cell circuitry contribute to neuronal dysfunction in ATXN1[82Q] mice. *Journal of Neuroscience*, **2011**, 31, 12778-89	6.6	61
97	Cerebellar Transcriptome Profiles of ATXN1 Transgenic Mice Reveal SCA1 Disease Progression and Protection Pathways. *Neuron*, **2016**, 89, 1194-1207	13.9	60

96	Organization of the human class I major histocompatibility complex genes. *Immunologic Research*, **1987**, 6, 1-10	4.3	58
95	Regional rescue of spinocerebellar ataxia type 1 phenotypes by 14-3-3epsilon haploinsufficiency in mice underscores complex pathogenicity in neurodegeneration. *Proceedings of the National Academy of Sciences of the United States of America*, **2011**, 108, 2142-7	11.5	56
94	Mapmodulin/leucine-rich acidic nuclear protein binds the light chain of microtubule-associated protein 1B and modulates neuritogenesis. *Journal of Biological Chemistry*, **2003**, 278, 34691-9	5.4	56
93	Antisense oligonucleotide-mediated ataxin-1 reduction prolongs survival in SCA1 mice and reveals disease-associated transcriptome profiles. *JCI Insight*, **2018**, 3,	9.9	54
92	Calcium dynamics and electrophysiological properties of cerebellar Purkinje cells in SCA1 transgenic mice. *Journal of Neurophysiology*, **2001**, 85, 1750-60	3.2	52
91	Progress in pathogenesis studies of spinocerebellar ataxia type 1. *Philosophical Transactions of the Royal Society B: Biological Sciences*, **1999**, 354, 1079-81	5.8	51
90	HLA non-A,B,C class I genes: their structure and expression. *Immunologic Research*, **1990**, 9, 265-74	4.3	50
89	Extensive cryptic splicing upon loss of RBM17 and TDP43 in neurodegeneration models. *Human Molecular Genetics*, **2016**, 25, 5083-5093	5.6	50
88	Molecular analysis of the variant alloantigen HLA-B27d (HLA-B*2703) identifies a unique single amino acid substitution. *Human Immunology*, **1988**, 21, 209-19	2.3	49
87	Partial loss of Tip60 slows mid-stage neurodegeneration in a spinocerebellar ataxia type 1 (SCA1) mouse model. *Human Molecular Genetics*, **2011**, 20, 2204-12	5.6	48
86	Phosphorylation of ATXN1 at Ser776 in the cerebellum. *Journal of Neurochemistry*, **2009**, 110, 675-86	6	47
85	Mapping of class I DNA sequences within the human major histocompatibility complex. *Immunogenetics*, **1983**, 18, 489-502	3.2	47
84	Cloning and developmental expression analysis of the murine homolog of the spinocerebellar ataxia type 1 gene (Sca1). *Human Molecular Genetics*, **1996**, 5, 33-40	5.6	46
83	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	13.9	45
82	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, 34606-16	5.4	43
81	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, 2789-95	5.6	43
80	Spinocerebellar ataxia type 1--modeling the pathogenesis of a polyglutamine neurodegenerative disorder in transgenic mice. *Journal of Neuropathology and Experimental Neurology*, **2000**, 59, 265-70	3.1	41
79	The effects of the polyglutamine repeat protein ataxin-1 on the UbL-UBA protein A1Up. *Journal of Biological Chemistry*, **2004**, 279, 42290-301	5.4	39

78	Purkinje cell ataxin-1 modulates climbing fiber synaptic input in developing and adult mouse cerebellum. *Journal of Neuroscience*, **2013**, 33, 5806-20	6.6	37
77	Reduction of Purkinje cell pathology in SCA1 transgenic mice by p53 deletion. *Neurobiology of Disease*, **2001**, 8, 974-81	7.5	35
76	Generation and characterization of LANP/pp32 null mice. *Molecular and Cellular Biology*, **2004**, 24, 3140-9	4.8	32
75	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, 5704-11	3.2	31
74	SCA1-phosphorylation, a regulator of Ataxin-1 function and pathogenesis. *Progress in Neurobiology*, **2012**, 99, 179-85	10.9	28
73	RNA targets of the fragile X protein. *Cell*, **2001**, 107, 555-7	56.2	28
72	A cell-based screen for modulators of ataxin-1 phosphorylation. *Human Molecular Genetics*, **2005**, 14, 1095-105	5.6	27
71	HLA-G transgenic mice: a model for studying expression and function at the maternal/fetal interface. *Immunological Reviews*, **1995**, 147, 53-65	11.3	26
70	The transcription factor E2F-1 in SV40 T antigen-induced cerebellar Purkinje cell degeneration. *Molecular and Cellular Neurosciences*, **1998**, 12, 16-28	4.8	25
69	FTD and ALS: genetic ties that bind. *Neuron*, **2011**, 72, 189-90	13.9	24
68	In vivo monitoring of recovery from neurodegeneration in conditional transgenic SCA1 mice. *Experimental Neurology*, **2011**, 232, 290-8	5.7	24
67	Susceptibility to cell death induced by mutant SV40 T-antigen correlates with Purkinje neuron functional development. *Molecular and Cellular Neurosciences*, **1997**, 9, 42-62	4.8	23
66	A native interactor scaffolds and stabilizes toxic ATAXIN-1 oligomers in SCA1. *ELife*, **2015**, 4,	8.9	23
65	Cytotoxic T lymphocyte recognition of HLA-G in mice. *Human Immunology*, **1997**, 55, 127-39	2.3	20
64	Protocol for genetic testing in Huntington disease: three years of experience in Minnesota. *American Journal of Medical Genetics Part A*, **1991**, 40, 518-22		20
63	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, 158-66	7.5	19
62	Polyglutamine disease toxicity is regulated by Nemo-like kinase in spinocerebellar ataxia type 1. *Journal of Neuroscience*, **2013**, 33, 9328-36	6.6	19
61	Mouse models of human CAG repeat disorders. *Brain Pathology*, **1997**, 7, 965-77	6	19

60	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	4.6	19
59	Genetically engineered mouse models of the trinucleotide-repeat spinocerebellar ataxias. *Brain Research Bulletin*, **2012**, 88, 33-42	3.9	18
58	Linkage of an Alzheimer disease susceptibility locus to markers on human chromosome 21. *American Journal of Medical Genetics Part A*, **1991**, 40, 449-53		18
57	Diminishing return for mechanistic therapeutics with neurodegenerative disease duration?: There may be a point in the course of a neurodegenerative condition where therapeutics targeting disease-causing mechanisms are futile. *BioEssays*, **2016**, 38, 977-80	4.1	18
56	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, 677-86	5.1	17
55	Spinocerebellar Ataxia Type 1: Molecular Mechanisms of Neurodegeneration and Preclinical Studies. *Advances in Experimental Medicine and Biology*, **2018**, 1049, 135-145	3.6	16
54	Diagnosis of classical steroid 21-hydroxylase deficiency using an HLA-B locus-specific DNA-probe. *American Journal of Medical Genetics Part A*, **1988**, 29, 703-12		16
53	Use of DNA probes from the 5′ flanking region of the HLA-B gene to examine polymorphism at the HLA-B locus. *Human Immunology*, **1986**, 16, 137-47	2.3	16
52	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	7.5	16
51	Characterization of the zebrafish atxn1/axh gene family. *Journal of Neurogenetics*, **2009**, 23, 313-23	1.6	15
50	The ins and outs of a polyglutamine neurodegenerative disease: spinocerebellar ataxia type 1 (SCA1). *Neurobiology of Disease*, **2000**, 7, 129-34	7.5	15
49	Targeting inhibitory cerebellar circuitry to alleviate behavioral deficits in a mouse model for studying idiopathic autism. *Neuropsychopharmacology*, **2020**, 45, 1159-1170	8.7	14
48	Motor neuron degeneration correlates with respiratory dysfunction in SCA1. *DMM Disease Models and Mechanisms*, **2018**, 11,	4.1	14
47	Identification of a novel phosphorylation site in ataxin-1. *Biochimica Et Biophysica Acta - Molecular Cell Research*, **2005**, 1744, 11-8	4.9	13
46	Microarrays and polyglutamine disorders: reports from the Hereditary Disease Array Group. *Human Molecular Genetics*, **2002**, 11, 1909-10	5.6	13
45	Overexpression of CREB reduces CRE-mediated transcription: behavioral and cellular analyses in transgenic mice. *Molecular and Cellular Neurosciences*, **2004**, 25, 602-11	4.8	12
44	Ataxin-1 oligomers induce local spread of pathology and decreasing them by passive immunization slows Spinocerebellar ataxia type 1 phenotypes. *ELife*, **2015**, 4,	8.9	12
43	PAK1 regulates ATXN1 levels providing an opportunity to modify its toxicity in spinocerebellar ataxia type 1. *Human Molecular Genetics*, **2018**, 27, 2863-2873	5.6	11

42	Hsp70/Hsc70 regulates the effect phosphorylation has on stabilizing ataxin-1. *Journal of Neurochemistry*, **2007**, 102, 2040-2048	6	11
41	Visualizing and mapping the cerebellum with serial optical coherence scanner. *Neurophotonics*, **2017**, 4, 011006	3.9	11
40	Regulation of ataxin-1 phosphorylation and its impact on biology. *Methods in Molecular Biology*, **2013**, 1010, 201-9	1.4	10
39	RNA gains a new function: a mediator of neurodegeneration. *Trends in Neurosciences*, **2004**, 27, 233-4	13.3	9
38	Qs in the nucleus. *Neuron*, **2001**, 31, 875-6	13.9	9
37	Unstable trinucleotide repeats and the diagnosis of neurodegenerative disease. *Human Pathology*, **1994**, 25, 598-601	3.7	9
36	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	20.1	9
35	HLA Class I Gene Family: Characterization of Genes Encoding Non-HLA-A,B,C Proteins **1989**, 33-40		9
34	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, 7533-4	11.5	8
33	Nuclear ataxias. *Cold Spring Harbor Perspectives in Biology*, **2010**, 2, a000786	10.2	8
32	Lurcher, nPIST, and autophagy. *Neuron*, **2002**, 35, 813-4	13.9	8
31	Pathogenesis of polyglutamine-induced disease: A model for SCA1. *Molecular Genetics and Metabolism*, **1999**, 66, 172-8	3.7	8
30	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, 723-34	4	7
29	Unstable nucleotide repeat minireview series: a molecular biography of unstable repeat disorders. *Journal of Biological Chemistry*, **2009**, 284, 7405	5.4	6
28	Polarization-sensitive optical coherence tomography reveals gray matter and white matter atrophy in SCA1 mouse models. *Neurobiology of Disease*, **2018**, 116, 69-77	7.5	6
27	The ataxin-1 interactome reveals direct connection with multiple disrupted nuclear transport pathways. *Nature Communications*, **2020**, 11, 3343	17.4	5
26	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	2.4	5
25	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	5.6	5

24	Into the depths of ataxia. *Journal of Clinical Investigation*, **2004**, 113, 505-7	15.9	4
23	Complete primary structure of human histocompatibility antigen HLA-B7: evolutionary and functional implications. *Current Topics in Developmental Biology*, **1980**, 14, 97-113	5.3	3
22	Antisense Oligonucleotide Therapeutic Approach for Suppression of Ataxin-1 Expression: A Safety Assessment. *Molecular Therapy - Nucleic Acids*, **2020**, 21, 1006-1016	10.7	3
21	Are polyglutamine diseases expanding?. *Neuron*, **2011**, 70, 377-8	13.9	2
20	Cholecystokinin 1 receptor activation restores normal mTORC1 signaling and is protective to Purkinje cells of SCA mice. *Cell Reports*, **2021**, 37, 109831	10.6	2
19	The Major Histocompatibility Complex: Analysis at the Protein and DNA Levels **1982**, 1-51		2
18	Modulation of ATXN1 S776 phosphorylation reveals the importance of allele-specific targeting in SCA1. *JCI Insight*, **2021**, 6,	9.9	2
17	Consensus Paper: Strengths and Weaknesses of Animal Models of Spinocerebellar Ataxias and Their Clinical Implications. *Cerebellum*, **2021**, 1	4.3	2
16	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
15	Altered Capicua expression drives regional Purkinje neuron vulnerability through ion channel gene dysregulation in Spinocerebellar ataxia type 1		1
14	Dual targeting of brain region-specific kinases potentiates neurological rescue in Spinocerebellar ataxia type 1. *EMBO Journal*, **2021**, 40, e106106	13	1
13	Complete Primary Structure of Human Histocompatibility Antigen HLA-B7 **1981**, 479-493		1
12	Autistic-like behavior and cerebellar dysfunction in Bmal1 mutant mice ameliorated by mTORC1 inhibition.. *Molecular Psychiatry*, **2022**,	15.1	1
11	Spinocerebellar Ataxias **2017**, 284-295		0
10	Stephen T. Warren, Ph.D. (1953-2021): A remembrance.. *American Journal of Human Genetics*, **2022**, 109, 3-11	11	0
9	Animal Models of Spinocerebellar Ataxia Type 1 **2015**, 979-990		
8	Animal Models of Spinocerebellar Ataxia Type 1 (SCA1) **2005**, 623-630		
7	Spinocerebellar Ataxia Type 1 **2006**, 87-99		

List of Publications

6 Spinocerebellar ataxia type 1 **2001**, 409-418

5 Spinocerebellar Ataxia 1 (SCA1) **2003**, 35-43

4 Phosphorylation of Ataxin-1: A Link Between Basic Research and Clinical Application in Spinocerebellar Ataxia Type 1 **2006**, 339-349

3 Spinocerebellar Ataxia Type 1 **2007**, 149-155

2 Transgenic Mouse Models of CAG Trinucleotide Repeat Neurologic Diseases **1999**, 163-185

1 Intrinsic Brain Signaling Pathways: Targets of Neuron Degeneration. *Research and Perspectives in Alzheimer's Disease*, **2011**, 125-131