Paul N Valdmanis

List of Publications by Year
in descending order

47 papers	6,124 citations	201674 27 h-index	214800 47 g-index
49 all docs	49 docs citations	49 times ranked	7644 citing authors

#	Article	IF	Citations
1	Mutations in the FUS/TLS Gene on Chromosome 16 Cause Familial Amyotrophic Lateral Sclerosis. Science, 2009, 323, 1205-1208.	12.6	2,302
2	TARDBP mutations in individuals with sporadic and familial amyotrophic lateral sclerosis. Nature Genetics, 2008, 40, 572-574.	21.4	1,371
3	The Loop Position of shRNAs and Pre-miRNAs Is Critical for the Accuracy of Dicer Processing InÂVivo. Cell, 2012, 151, 900-911.	28.9	266
4	Promoterless gene targeting without nucleases ameliorates haemophilia B in mice. Nature, 2015, 517, 360-364.	27.8	226
5	Genetics of familial amyotrophic lateral sclerosis. Neurology, 2008, 70, 144-152.	1.1	189
6	Mutations in the KIAA0196 Gene at the SPG8 Locus Cause Hereditary Spastic Paraplegia. American Journal of Human Genetics, 2007, 80, 152-161.	6.2	168
7	Contribution of TARDBP mutations to sporadic amyotrophic lateral sclerosis. Journal of Medical Genetics, 2008, 46, 112-114.	3.2	162
8	Oxidized/misfolded superoxide dismutaseâ€1: the cause of all amyotrophic lateral sclerosis?. Annals of Neurology, 2007, 62, 553-559.	5.3	137
9	From animal models to human disease: a genetic approach for personalized medicine in ALS. Acta Neuropathologica Communications, 2016, 4, 70.	5.2	115
10	Three Families With Amyotrophic Lateral Sclerosis and Frontotemporal Dementia With Evidence of Linkage to Chromosome 9p. Archives of Neurology, 2007, 64, 240.	4.5	111
11	Recent advances in the genetics of amyotrophic lateral sclerosis. Current Neurology and Neuroscience Reports, 2009, 9, 198-205.	4.2	103
12	Investigating the contribution of VAPB/ALS8 loss of function in amyotrophic lateral sclerosis. Human Molecular Genetics, 2013, 22, 2350-2360.	2.9	75
13	Integrative geneâ€“tissue microarray-based approach for identification of human disease biomarkers: application to amyotrophic lateral sclerosis. Human Molecular Genetics, 2010, 19, 3233-3253.	2.9	58
14	Four familial ALS pedigrees discordant for two SOD1 mutations: are all SOD1 mutations pathogenic?. Journal of Neurology, Neurosurgery and Psychiatry, 2010, 81, 572-577.	1.9	57
15	Resequencing of 29 Candidate Genes in Patients With Familial and Sporadic Amyotrophic Lateral Sclerosis. Archives of Neurology, 2011, 68, 587-93.	4.5	52
16	A tRNA-Derived Small RNA Regulates Ribosomal Protein S28 Protein Levels after Translation Initiation in Humans and Mice. Cell Reports, 2019, 29, 3816-3824.e4.	6.4	52
17	miR-122 removal in the liver activates imprinted microRNAs and enables more effective microRNA-mediated gene repression. Nature Communications, 2018, 9, 5321.	12.8	48
18	Upregulation of the microRNA cluster at the Dlk1-Dio3 locus in lung adenocarcinoma. Oncogene, 2015, 34, 94-103.	5.9	46

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19	Future of rAAV Gene Therapy: Platform for RNAi, Gene Editing, and Beyond. Human Gene Therapy, 2017, 28, 361-372.	2.7	40
20	Evolution of a Human-Specific Tandem Repeat Associated with ALS. American Journal of Human Genetics, 2020, 107, 445-460.	6.2	39
21	Expression determinants of mammalian argonaute proteins in mediating gene silencing. Nucleic Acids Research, 2012, 40, 3704-3713.	14.5	35
22	rAAV-Mediated Tumorigenesis: Still Unresolved After an AAV Assault. Molecular Therapy, 2012, 20, 2014-2017.	8.2	33
23	A Mutation that Creates a Pseudoexon in SOD1 Causes Familial ALS. Annals of Human Genetics, 2009, 73, 652-657.	0.8	32
24	A mutation in the RNF170 gene causes autosomal dominant sensory ataxia. Brain, 2011, 134, 602-607.	7.6	32
25	RNA interferenceâ€“induced hepatotoxicity results from loss of the first synthesized isoform of microRNA-122 in mice. Nature Medicine, 2016, 22, 557-562.	30.7	32
26	50bp deletion in the promoter for superoxide dismutase 1 (SOD1) reduces SOD1 expression in vitro and may correlate with increased age of onset of sporadic amyotrophic lateral sclerosis. Amyotrophic Lateral Sclerosis and Other Motor Neuron Disorders, 2008, 9, 229-237.	2.1	29
27	Alternative splicing in a presenilin 2 variant associated with Alzheimer disease. Annals of Clinical and Translational Neurology, 2019, 6, 762-777.	3.7	29
28	Autosomal dominant primary lateral sclerosis. Neurology, 2007, 68, 1156-1157.	1.1	28
29	The Expanding Repertoire of Circular RNAs. Molecular Therapy, 2013, 21, 1112-1114.	8.2	28
30	Regulation of microRNA-mediated gene silencing by microRNA precursors. Nature Structural and Molecular Biology, 2014, 21, 825-832.	8.2	23
31	The proportion of mutations predicted to have a deleterious effect differs between gain and loss of function genes in neurodegenerative disease. Human Mutation, 2009, 30, E481-E489.	2.5	20
32	Tracking Adeno-Associated Virus Capsid Evolution by High-Throughput Sequencing. Human Gene Therapy, 2020, 31, 553-564.	2.7	19
33	A novel TYMP mutation in a French Canadian patient with mitochondrial neurogastrointestinal encephalomyopathy. Clinical Neurology and Neurosurgery, 2009, 111, 691-694.	1.4	18
34	Analysis ofDPP6andFGGYas candidate genes for amyotrophic lateral sclerosis. Amyotrophic Lateral Sclerosis and Other Motor Neuron Disorders, 2010, 11, 389-391.	2.1	18
35	Characterizing nucleotide variation and expansion dynamics in human-specific variable number tandem repeats. Genome Research, 2021, 31, 1313-1324.	5.5	15
36	Progress in Amyotrophic Lateral Sclerosis Gene Discovery. Neurology: Genetics, 2022, 8, .	1.9	15

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37	Human Induced Pluripotent Stem Cell-Derived TDP-43 Mutant Neurons Exhibit Consistent Functional Phenotypes Across Multiple Gene Edited Lines Despite Transcriptomic and Splicing Discrepancies. Frontiers in Cell and Developmental Biology, 2021, 9, 728707.	3.7	13
38	Autosomal dominant sensory ataxia: a neuroaxonal dystrophy. Acta Neuropathologica, 2008, 116, 331-336.	7.7	12
39	A Novel Mutation in a Large French-Canadian Family with LGMD1B. Canadian Journal of Neurological Sciences, 2008, 35, 331-334.	0.5	12
40	A Locus for Primary Lateral Sclerosis on Chromosome 4ptel-4p16.1. Archives of Neurology, 2008, 65, 383-6.	4.5	12
41	No TARDBP Mutations in a French Canadian Population of Patients With Parkinson Disease. Archives of Neurology, 2009, 66, 281-2.	4.5	12
42	Prophylactic In Vivo Hematopoietic Stem Cell Gene Therapy with an Immune Checkpoint Inhibitor Reverses Tumor Growth in Syngeneic Mouse Tumor Models. Cancer Research, 2020, 80, 549-560.	0.9	12
43	ALS predisposition modifiers: Knock NOX, who's there? SOD1 mice still are. European Journal of Human Genetics, 2008, 16, 140-142.	2.8	11
44	Expression of an alternatively spliced variant of SORL1 in neuronal dendrites is decreased in patients with Alzheimerâ€™s disease. Acta Neuropathologica Communications, 2021, 9, 43.	5.2	7
45	Endogenous MicroRNA Competition as a Mechanism of shRNA-Induced Cardiotoxicity. Molecular Therapy - Nucleic Acids, 2020, 19, 572-580.	5.1	4
46	Amyotrophic Lateral Sclerosis: Genome-wide association studies in amyotrophic lateral sclerosis. European Journal of Human Genetics, 2009, 17, 137-138.	2.8	3
47	A Complete Pipeline for Isolating and Sequencing MicroRNAs, and Analyzing Them Using Open Source Tools. Journal of Visualized Experiments, 2019, , .	0.3	2