Nicholas J Maragakis

List of Publications by Year
in descending order

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87 papers	8,706 citations	66315 42 h-index	54882 84 g-index
91 all docs	91 docs citations	91 times ranked	10927 citing authors

#	Article	IF	Citations
1	RNA Toxicity from the ALS/FTD C9ORF72 Expansion Is Mitigated by Antisense Intervention. Neuron, 2013, 80, 415-428.	3.8	785
2	C9orf72 nucleotide repeat structures initiate molecular cascades of disease. Nature, 2014, 507, 195-200.	13.7	779
3	Focal loss of the glutamate transporter EAAT2 in a transgenic rat model of SOD1 mutant-mediated amyotrophic lateral sclerosis (ALS). Proceedings of the National Academy of Sciences of the United States of America, 2002, 99, 1604-1609.	3.3	766
4	Mechanisms of Disease: astrocytes in neurodegenerative disease. Nature Clinical Practice Neurology, 2006, 2, 679-689.	2.7	700
5	Genome-wide Analyses Identify KIF5A as a Novel ALS Gene. Neuron, 2018, 97, 1268-1283.e6.	3.8	517
6	Protective Role of Reactive Astrocytes in Brain Ischemia. Journal of Cerebral Blood Flow and Metabolism, 2008, 28, 468-481.	2.4	441
7	Focal transplantationâ€“based astrocyte replacement is neuroprotective in a model of motor neuron disease. Nature Neuroscience, 2008, 11, 1294-1301.	7.1	403
8	Glutamate transporters: animal models to neurologic disease. Neurobiology of Disease, 2004, 15, 461-473.	2.1	321
9	Recovery from paralysis in adult rats using embryonic stem cells. Annals of Neurology, 2006, 60, 32-44.	2.8	266
10	Human Embryonic Germ Cell Derivatives Facilitate Motor Recovery of Rats with Diffuse Motor Neuron Injury. Journal of Neuroscience, 2003, 23, 5131-5140.	1.7	239
11	Astrocytes carrying the superoxide dismutase 1 (SOD1 ^{G93A}) mutation induce wild-type motor neuron degeneration in vivo. Proceedings of the National Academy of Sciences of the United States of America, 2011, 108, 17803-17808.	3.3	194
12	Concordant but Varied Phenotypes among Duchenne Muscular Dystrophy Patient-Specific Myoblasts Derived using a Human iPSC-Based Model. Cell Reports, 2016, 15, 2301-2312.	2.9	141
13	Electrical impedance myography as a biomarker to assess ALS progression. Amyotrophic Lateral Sclerosis and Other Motor Neuron Disorders, 2012, 13, 439-445.	2.3	125
14	Altered expression of the glutamate transporter EAAT2b in neurological disease. Annals of Neurology, 2004, 55, 469-477.	2.8	122
15	Connexin 43 in astrocytes contributes to motor neuron toxicity in amyotrophic lateral sclerosis. Glia, 2016, 64, 1154-1169.	2.5	114
16	Loss of the astrocyte glutamate transporter GLT1 modifies disease in SOD1G93A mice. Experimental Neurology, 2006, 201, 120-130.	2.0	113
17	Intraparenchymal spinal cord delivery of adeno-associated virus IGF-1 is protective in the SOD1G93A model of ALS. Brain Research, 2007, 1185, 256-265.	1.1	112
18	Human Glial-Restricted Progenitor Transplantation into Cervical Spinal Cord of the SOD1G93A Mouse Model of ALS. PLoS ONE, 2011, 6, e25968.	1.1	107

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19	Sporadic ALS Astrocytes Induce Neuronal Degeneration InÂVivo. Stem Cell Reports, 2017, 8, 843-855.	2.3	105
20	Viral-Induced Spinal Motor Neuron Death Is Non-Cell-Autonomous and Involves Glutamate Excitotoxicity. Journal of Neuroscience, 2004, 24, 7566-7575.	1.7	96
21	Multimodal Actions of Neural Stem Cells in a Mouse Model of ALS: A Meta-Analysis. Science Translational Medicine, 2012, 4, 165ra164.	5.8	91
22	Adult glial precursor proliferation in mutant SOD1^{G93A} mice. Glia, 2008, 56, 200-208.	2.5	81
23	Effect of Ezogabine on Cortical and Spinal Motor Neuron Excitability in Amyotrophic Lateral Sclerosis. JAMA Neurology, 2021, 78, 186.	4.5	79
24	Selective ablation of proliferating astrocytes does not affect disease outcome in either acute or chronic models of motor neuron degeneration. Experimental Neurology, 2008, 211, 423-432.	2.0	77
25	Glutamate transporter expression and function in human glial progenitors. Glia, 2004, 45, 133-143.	2.5	74
26	Defining SOD1 ALS natural history to guide therapeutic clinical trial design. Journal of Neurology, Neurosurgery and Psychiatry, 2017, 88, 99-105.	0.9	68
27	MicroRNA Profiling Reveals Marker of Motor Neuron Disease in ALS Models. Journal of Neuroscience, 2017, 37, 5574-5586.	1.7	66
28	Answer ALS, a large-scale resource for sporadic and familial ALS combining clinical and multi-omics data from induced pluripotent cell lines. Nature Neuroscience, 2022, 25, 226-237.	7.1	66
29	Degeneration of respiratory motor neurons in the SOD1 G93A transgenic rat model of ALS. Neurobiology of Disease, 2006, 21, 110-118.	2.1	63
30	Electrical impedance myography correlates with standard measures of Als severity. Muscle and Nerve, 2014, 49, 441-443.	1.0	61
31	Human iPS cell-derived astrocyte transplants preserve respiratory function after spinal cord injury. Experimental Neurology, 2015, 271, 479-492.	2.0	61
32	Addressing heterogeneity in amyotrophic lateral sclerosis CLINICAL TRIALS. Muscle and Nerve, 2020, 62, 156-166.	1.0	60
33	Induced pluripotent stem cells from ALS patients for disease modeling. Brain Research, 2015, 1607, 15-25.	1.1	57
34	Astrocytes drive upregulation of the multidrug resistance transporter ABCB1 (Pâ€Glycoprotein) in endothelial cells of the bloodâ€“brain barrier in mutant superoxide dismutase 1â€linked amyotrophic lateral sclerosis. Glia, 2016, 64, 1298-1313.	2.5	57
35	Sciatic nerve tumor and tumor-like lesionsâ€”uncommon pathologies. Skeletal Radiology, 2012, 41, 763-774.	1.2	55
36	A stocked toolbox for understanding the role of astrocytes in disease. Nature Reviews Neurology, 2018, 14, 351-362.	4.9	53

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37	Motor neuron-derived microRNAs cause astrocyte dysfunction in amyotrophic lateral sclerosis. Brain, 2018, 141, 2561-2575.	3.7	50
38	Altered astrocytic expression of TDP-43 does not influence motor neuron survival. Experimental Neurology, 2013, 250, 250-259.	2.0	49
39	Fibroblast bioenergetics to classify amyotrophic lateral sclerosis patients. Molecular Neurodegeneration, 2017, 12, 76.	4.4	49
40	Topiramate protects against motor neuron degeneration in organotypic spinal cord cultures but not in G93A SOD1 transgenic mice. Neuroscience Letters, 2003, 338, 107-110.	1.0	48
41	Reduction in expression of the astrocyte glutamate transporter, GLT1, worsens functional and histological outcomes following traumatic spinal cord injury. Glia, 2011, 59, 1996-2005.	2.5	48
42	Association of Variants in the SPTLC1 Gene With Juvenile Amyotrophic Lateral Sclerosis. JAMA Neurology, 2021, 78, 1236.	4.5	46
43	Advances in stem cell research for Amyotrophic Lateral Sclerosis. Current Opinion in Biotechnology, 2009, 20, 545-551.	3.3	45
44	A Comprehensive Library of Familial Human Amyotrophic Lateral Sclerosis Induced Pluripotent Stem Cells. PLoS ONE, 2015, 10, e0118266.	1.1	45
45	A randomized controlled trial of resistance and endurance exercise in amyotrophic lateral sclerosis. Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration, 2018, 19, 250-258.	1.1	44
46	Amyotrophic lateral sclerosis care and research in the United States during the <scp>COVID</scp>â€19 pandemic: Challenges and opportunities. Muscle and Nerve, 2020, 62, 182-186.	1.0	42
47	Glial restricted precursors protect against chronic glutamate neurotoxicity of motor neurons in vitro. Glia, 2005, 50, 145-159.	2.5	40
48	Neural and glial progenitor transplantation as a neuroprotective strategy for Amyotrophic Lateral Sclerosis (ALS). Brain Research, 2015, 1628, 343-350.	1.1	39
49	Multi-omic analysis of selectively vulnerable motor neuron subtypes implicates altered lipid metabolism in ALS. Nature Neuroscience, 2021, 24, 1673-1685.	7.1	38
50	Gene Profiling of Human Induced Pluripotent Stem Cell-Derived Astrocyte Progenitors Following Spinal Cord Engraftment. Stem Cells Translational Medicine, 2014, 3, 575-585.	1.6	37
51	Peripheral hyperstimulation alters site of disease onset and course in SOD1 rats. Neurobiology of Disease, 2010, 39, 252-264.	2.1	36
52	Spatial and temporal changes in promoter activity of the astrocyte glutamate transporter GLT1 following traumatic spinal cord injury. Journal of Neuroscience Research, 2011, 89, 1001-1017.	1.3	35
53	Transplantation of Glial Progenitors That Overexpress Glutamate Transporter GLT1 Preserves Diaphragm Function Following Cervical SCI. Molecular Therapy, 2015, 23, 533-548.	3.7	35
54	Role of Human-Induced Pluripotent Stem Cell-Derived Spinal Cord Astrocytes in the Functional Maturation of Motor Neurons in a Multielectrode Array System. Stem Cells Translational Medicine, 2019, 8, 1272-1285.	1.6	34

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55	Cx43 hemichannels contribute to astrocyte-mediated toxicity in sporadic and familial ALS. Proceedings of the National Academy of Sciences of the United States of America, 2022, 119, e2107391119.	3.3	29
56	Olfactory dysfunction in amyotrophic lateral sclerosis. Annals of Clinical and Translational Neurology, 2018, 5, 976-981.	1.7	28
57	Generation of <scp>GFAP::GFP</scp> astrocyte reporter lines from human adult fibroblastâ€derived i<scp>PS</scp> cells using zincâ€finger nuclease technology. Glia, 2016, 64, 63-75.	2.5	26
58	Preâ€morbid type 2 diabetes mellitus is not a prognostic factor in amyotrophic lateral sclerosis. Muscle and Nerve, 2015, 52, 339-343.	1.0	25
59	Glia: an emerging target for neurological disease therapy. Stem Cell Research and Therapy, 2012, 3, 37.	2.4	24
60	Human glial progenitor engraftment and gene expression is independent of the ALS environment. Experimental Neurology, 2015, 264, 188-199.	2.0	21
61	Mini-Review: Induced pluripotent stem cells and the search for new cell-specific ALS therapeutic targets. Neuroscience Letters, 2021, 755, 135911.	1.0	20
62	Impaired SDF1/CXCR4 signaling in glial progenitors derived from SOD1G93A mice. Journal of Neuroscience Research, 2007, 85, 2422-2432.	1.3	19
63	Current and emerging ALS biomarkers: utility and potential in clinical trials. Expert Review of Neurotherapeutics, 2018, 18, 871-886.	1.4	18
64	Primary Lateral Sclerosis and Early Upper Motor Neuron Disease. Journal of Clinical Neuromuscular Disease, 2016, 17, 99-105.	0.3	17
65	Primary lateral sclerosis (PLS) functional rating scale: PLSâ€specific clinimetric scale. Muscle and Nerve, 2020, 61, 163-172.	1.0	17
66	Expression and Cellular Distribution of P-Glycoprotein and Breast Cancer Resistance Protein in Amyotrophic Lateral Sclerosis Patients. Journal of Neuropathology and Experimental Neurology, 2020, 79, 266-276.	0.9	17
67	A Spontaneous Missense Mutation in Branched Chain Keto Acid Dehydrogenase Kinase in the Rat Affects Both the Central and Peripheral Nervous Systems. PLoS ONE, 2016, 11, e0160447.	1.1	16
68	MN-166 (ibudilast) in amyotrophic lateral sclerosis in a Phase IIb/III study: COMBAT-ALS study design. Neurodegenerative Disease Management, 2021, 11, 431-443.	1.2	16
69	Stem cells and the ALS neurologist. Amyotrophic Lateral Sclerosis and Other Motor Neuron Disorders, 2010, 11, 417-423.	2.3	15
70	What can we learn from the edaravone development program for ALS?. Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration, 2017, 18, 98-103.	1.1	15
71	Amyotrophic Lateral Sclerosis: Pathogenesis, Differential Diagnoses, And Potential Interventions. Journal of Spinal Cord Medicine, 2002, 25, 262-273.	0.7	12
72	Serial in vivo imaging of transplanted allogeneic neural stem cell survival in a mouse model of amyotrophic lateral sclerosis. Experimental Neurology, 2017, 289, 96-102.	2.0	11

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73	Perfluorocarbon Labeling of Human Glial-Restricted Progenitors for 19F Magnetic Resonance Imaging. Stem Cells Translational Medicine, 2019, 8, 355-365.	1.6	11
74	Edaravone activates the GDNF/RET neurotrophic signaling pathway and protects mRNA-induced motor neurons from iPS cells. Molecular Neurodegeneration, 2022, 17, 8.	4.4	10
75	Focal and dose-dependent neuroprotection in ALS mice following AAV2-neurturin delivery. Experimental Neurology, 2020, 323, 113091.	2.0	9
76	Safety and efficacy of oral levosimendan in people with amyotrophic lateral sclerosis (the REFALS) Tj ETQq0 0 0 rgBT /Overlock 10 Tf 50 821-831.	4.9	9
77	Hemiparetic Primary Lateral Sclerosis: Revisiting Mills Syndrome. Case Reports in Neurology, 2015, 7, 191-195.	0.3	7
78	Stem Cell Transplantation for Spinal Cord Neurodegeneration. Methods in Molecular Biology, 2011, 793, 479-493.	0.4	6
79	Exploring Motor Neuron Diseases Using iPSC Platforms. Stem Cells, 2022, 40, 2-13.	1.4	6
80	Motoneuron Disease: Basic Science. Advances in Neurobiology, 2017, 15, 163-190.	1.3	5
81	Analyzing progression of motor and speech impairment in ALS. , 2019, 2019, 6097-6102.		5
82	Establishment of an Electrophysiological Platform for Modeling ALS with Regionally-Specific Human Pluripotent Stem Cell-Derived Astrocytes and Neurons. Journal of Visualized Experiments, 2021, , .	0.2	5
83	Motoneuron Disease: Clinical. Advances in Neurobiology, 2017, 15, 191-210.	1.3	2
84	Rethinking a drug treatment failure on a traditional ALS target. Experimental Neurology, 2009, 216, 254-257.	2.0	1
85	New Treatments in Amyotrophic Lateral Sclerosis. Neuropsychopharmacology, 2011, 36, 370-372.	2.8	1
86	Erratum. Advances in Neurobiology, 2017, 15, E1-E1.	1.3	1
87	Human induced pluripotent stem cellâ€“derived astrocytes progenitors as discovery platforms. , 2022, , 45-89.		0