

Nicholas J Maragakis

List of Publications by Year in descending order

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87
papers

8,706
citations

66343

42
h-index

54911

84
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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. <i>Neuron</i> , 2013, 80, 415-428.	8.1	785
2	C9orf72 nucleotide repeat structures initiate molecular cascades of disease. <i>Nature</i> , 2014, 507, 195-200.	27.8	779
3	Focal loss of the glutamate transporter EAAT2 in a transgenic rat model of SOD1 mutant-mediated amyotrophic lateral sclerosis (ALS). <i>Proceedings of the National Academy of Sciences of the United States of America</i> , 2002, 99, 1604-1609.	7.1	766
4	Mechanisms of Disease: astrocytes in neurodegenerative disease. <i>Nature Clinical Practice Neurology</i> , 2006, 2, 679-689.	2.5	700
5	Genome-wide Analyses Identify KIF5A as a Novel ALS Gene. <i>Neuron</i> , 2018, 97, 1268-1283.e6.	8.1	517
6	Protective Role of Reactive Astrocytes in Brain Ischemia. <i>Journal of Cerebral Blood Flow and Metabolism</i> , 2008, 28, 468-481.	4.3	441
7	Focal transplantation-based astrocyte replacement is neuroprotective in a model of motor neuron disease. <i>Nature Neuroscience</i> , 2008, 11, 1294-1301.	14.8	403
8	Glutamate transporters: animal models to neurologic disease. <i>Neurobiology of Disease</i> , 2004, 15, 461-473.	4.4	321
9	Recovery from paralysis in adult rats using embryonic stem cells. <i>Annals of Neurology</i> , 2006, 60, 32-44.	5.3	266
10	Human Embryonic Germ Cell Derivatives Facilitate Motor Recovery of Rats with Diffuse Motor Neuron Injury. <i>Journal of Neuroscience</i> , 2003, 23, 5131-5140.	3.6	239
11	Astrocytes carrying the superoxide dismutase 1 (SOD1 ^{G93A}) mutation induce wild-type motor neuron degeneration in vivo. <i>Proceedings of the National Academy of Sciences of the United States of America</i> , 2011, 108, 17803-17808.	7.1	194
12	Concordant but Varied Phenotypes among Duchenne Muscular Dystrophy Patient-Specific Myoblasts Derived using a Human iPSC-Based Model. <i>Cell Reports</i> , 2016, 15, 2301-2312.	6.4	141
13	Electrical impedance myography as a biomarker to assess ALS progression. <i>Amyotrophic Lateral Sclerosis and Other Motor Neuron Disorders</i> , 2012, 13, 439-445.	2.1	125
14	Altered expression of the glutamate transporter EAAT2b in neurological disease. <i>Annals of Neurology</i> , 2004, 55, 469-477.	5.3	122
15	Connexin 43 in astrocytes contributes to motor neuron toxicity in amyotrophic lateral sclerosis. <i>Glia</i> , 2016, 64, 1154-1169.	4.9	114
16	Loss of the astrocyte glutamate transporter GLT1 modifies disease in SOD1G93A mice. <i>Experimental Neurology</i> , 2006, 201, 120-130.	4.1	113
17	Intraparenchymal spinal cord delivery of adeno-associated virus IGF-1 is protective in the SOD1G93A model of ALS. <i>Brain Research</i> , 2007, 1185, 256-265.	2.2	112
18	Human Glial-Restricted Progenitor Transplantation into Cervical Spinal Cord of the SOD1G93A Mouse Model of ALS. <i>PLoS ONE</i> , 2011, 6, e25968.	2.5	107

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19	Sporadic ALS Astrocytes Induce Neuronal Degeneration In Vivo. <i>Stem Cell Reports</i> , 2017, 8, 843-855.	4.8	105
20	Viral-Induced Spinal Motor Neuron Death Is Non-Cell-Autonomous and Involves Glutamate Excitotoxicity. <i>Journal of Neuroscience</i> , 2004, 24, 7566-7575.	3.6	96
21	Multimodal Actions of Neural Stem Cells in a Mouse Model of ALS: A Meta-Analysis. <i>Science Translational Medicine</i> , 2012, 4, 165ra164.	12.4	91
22	Adult glial precursor proliferation in mutant SOD1 ^{G93A} mice. <i>Glia</i> , 2008, 56, 200-208.	4.9	81
23	Effect of Ezogabine on Cortical and Spinal Motor Neuron Excitability in Amyotrophic Lateral Sclerosis. <i>JAMA Neurology</i> , 2021, 78, 186.	9.0	79
24	Selective ablation of proliferating astrocytes does not affect disease outcome in either acute or chronic models of motor neuron degeneration. <i>Experimental Neurology</i> , 2008, 211, 423-432.	4.1	77
25	Glutamate transporter expression and function in human glial progenitors. <i>Glia</i> , 2004, 45, 133-143.	4.9	74
26	Defining SOD1 ALS natural history to guide therapeutic clinical trial design. <i>Journal of Neurology, Neurosurgery and Psychiatry</i> , 2017, 88, 99-105.	1.9	68
27	MicroRNA Profiling Reveals Marker of Motor Neuron Disease in ALS Models. <i>Journal of Neuroscience</i> , 2017, 37, 5574-5586.	3.6	66
28	Answer ALS, a large-scale resource for sporadic and familial ALS combining clinical and multi-omics data from induced pluripotent cell lines. <i>Nature Neuroscience</i> , 2022, 25, 226-237.	14.8	66
29	Degeneration of respiratory motor neurons in the SOD1 G93A transgenic rat model of ALS. <i>Neurobiology of Disease</i> , 2006, 21, 110-118.	4.4	63
30	Electrical impedance myography correlates with standard measures of ALS severity. <i>Muscle and Nerve</i> , 2014, 49, 441-443.	2.2	61
31	Human iPS cell-derived astrocyte transplants preserve respiratory function after spinal cord injury. <i>Experimental Neurology</i> , 2015, 271, 479-492.	4.1	61
32	Addressing heterogeneity in amyotrophic lateral sclerosis CLINICAL TRIALS. <i>Muscle and Nerve</i> , 2020, 62, 156-166.	2.2	60
33	Induced pluripotent stem cells from ALS patients for disease modeling. <i>Brain Research</i> , 2015, 1607, 15-25.	2.2	57
34	Astrocytes drive upregulation of the multidrug resistance transporter ABCB1 (P-glycoprotein) in endothelial cells of the blood-brain barrier in mutant superoxide dismutase 1-linked amyotrophic lateral sclerosis. <i>Glia</i> , 2016, 64, 1298-1313.	4.9	57
35	Sciatic nerve tumor and tumor-like lesions—uncommon pathologies. <i>Skeletal Radiology</i> , 2012, 41, 763-774.	2.0	55
36	A stocked toolbox for understanding the role of astrocytes in disease. <i>Nature Reviews Neurology</i> , 2018, 14, 351-362.	10.1	53

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37	Motor neuron-derived microRNAs cause astrocyte dysfunction in amyotrophic lateral sclerosis. <i>Brain</i> , 2018, 141, 2561-2575.	7.6	50
38	Altered astrocytic expression of TDP-43 does not influence motor neuron survival. <i>Experimental Neurology</i> , 2013, 250, 250-259.	4.1	49
39	Fibroblast bioenergetics to classify amyotrophic lateral sclerosis patients. <i>Molecular Neurodegeneration</i> , 2017, 12, 76.	10.8	49
40	Topiramate protects against motor neuron degeneration in organotypic spinal cord cultures but not in G93A SOD1 transgenic mice. <i>Neuroscience Letters</i> , 2003, 338, 107-110.	2.1	48
41	Reduction in expression of the astrocyte glutamate transporter, GLT1, worsens functional and histological outcomes following traumatic spinal cord injury. <i>Glia</i> , 2011, 59, 1996-2005.	4.9	48
42	Association of Variants in the <i>SPTLC1</i> Gene With Juvenile Amyotrophic Lateral Sclerosis. <i>JAMA Neurology</i> , 2021, 78, 1236.	9.0	46
43	Advances in stem cell research for Amyotrophic Lateral Sclerosis. <i>Current Opinion in Biotechnology</i> , 2009, 20, 545-551.	6.6	45
44	A Comprehensive Library of Familial Human Amyotrophic Lateral Sclerosis Induced Pluripotent Stem Cells. <i>PLoS ONE</i> , 2015, 10, e0118266.	2.5	45
45	A randomized controlled trial of resistance and endurance exercise in amyotrophic lateral sclerosis. <i>Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration</i> , 2018, 19, 250-258.	1.7	44
46	Amyotrophic lateral sclerosis care and research in the United States during the COVID-19 pandemic: Challenges and opportunities. <i>Muscle and Nerve</i> , 2020, 62, 182-186.	2.2	42
47	Glial restricted precursors protect against chronic glutamate neurotoxicity of motor neurons in vitro. <i>Glia</i> , 2005, 50, 145-159.	4.9	40
48	Neural and glial progenitor transplantation as a neuroprotective strategy for Amyotrophic Lateral Sclerosis (ALS). <i>Brain Research</i> , 2015, 1628, 343-350.	2.2	39
49	Multi-omic analysis of selectively vulnerable motor neuron subtypes implicates altered lipid metabolism in ALS. <i>Nature Neuroscience</i> , 2021, 24, 1673-1685.	14.8	38
50	Gene Profiling of Human Induced Pluripotent Stem Cell-Derived Astrocyte Progenitors Following Spinal Cord Engraftment. <i>Stem Cells Translational Medicine</i> , 2014, 3, 575-585.	3.3	37
51	Peripheral hyperstimulation alters site of disease onset and course in SOD1 rats. <i>Neurobiology of Disease</i> , 2010, 39, 252-264.	4.4	36
52	Spatial and temporal changes in promoter activity of the astrocyte glutamate transporter GLT1 following traumatic spinal cord injury. <i>Journal of Neuroscience Research</i> , 2011, 89, 1001-1017.	2.9	35
53	Transplantation of Glial Progenitors That Overexpress Glutamate Transporter GLT1 Preserves Diaphragm Function Following Cervical SCI. <i>Molecular Therapy</i> , 2015, 23, 533-548.	8.2	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. <i>Stem Cells Translational Medicine</i> , 2019, 8, 1272-1285.	3.3	34

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55	Cx43 hemichannels contribute to astrocyte-mediated toxicity in sporadic and familial ALS. <i>Proceedings of the National Academy of Sciences of the United States of America</i> , 2022, 119, e2107391119.	7.1	29
56	Olfactory dysfunction in amyotrophic lateral sclerosis. <i>Annals of Clinical and Translational Neurology</i> , 2018, 5, 976-981.	3.7	28
57	Generation of <sc>GFAP::GFP</sc> astrocyte reporter lines from human adult fibroblast-derived i<sc>PS</sc> cells using zinc-finger nuclease technology. <i>Glia</i> , 2016, 64, 63-75.	4.9	26
58	Pre-morbid type 2 diabetes mellitus is not a prognostic factor in amyotrophic lateral sclerosis. <i>Muscle and Nerve</i> , 2015, 52, 339-343.	2.2	25
59	Glia: an emerging target for neurological disease therapy. <i>Stem Cell Research and Therapy</i> , 2012, 3, 37.	5.5	24
60	Human glial progenitor engraftment and gene expression is independent of the ALS environment. <i>Experimental Neurology</i> , 2015, 264, 188-199.	4.1	21
61	Mini-Review: Induced pluripotent stem cells and the search for new cell-specific ALS therapeutic targets. <i>Neuroscience Letters</i> , 2021, 755, 135911.	2.1	20
62	Impaired SDF1/CXCR4 signaling in glial progenitors derived from SOD1G93A mice. <i>Journal of Neuroscience Research</i> , 2007, 85, 2422-2432.	2.9	19
63	Current and emerging ALS biomarkers: utility and potential in clinical trials. <i>Expert Review of Neurotherapeutics</i> , 2018, 18, 871-886.	2.8	18
64	Primary Lateral Sclerosis and Early Upper Motor Neuron Disease. <i>Journal of Clinical Neuromuscular Disease</i> , 2016, 17, 99-105.	0.7	17
65	Primary lateral sclerosis (PLS) functional rating scale: PLS-specific clinimetric scale. <i>Muscle and Nerve</i> , 2020, 61, 163-172.	2.2	17
66	Expression and Cellular Distribution of P-Glycoprotein and Breast Cancer Resistance Protein in Amyotrophic Lateral Sclerosis Patients. <i>Journal of Neuropathology and Experimental Neurology</i> , 2020, 79, 266-276.	1.7	17
67	A Spontaneous Missense Mutation in Branched Chain Keto Acid Dehydrogenase Kinase in the Rat Affects Both the Central and Peripheral Nervous Systems. <i>PLoS ONE</i> , 2016, 11, e0160447.	2.5	16
68	MN-166 (ibudilast) in amyotrophic lateral sclerosis in a Phase IIb/III study: COMBAT-ALS study design. <i>Neurodegenerative Disease Management</i> , 2021, 11, 431-443.	2.2	16
69	Stem cells and the ALS neurologist. <i>Amyotrophic Lateral Sclerosis and Other Motor Neuron Disorders</i> , 2010, 11, 417-423.	2.1	15
70	What can we learn from the edaravone development program for ALS?. <i>Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration</i> , 2017, 18, 98-103.	1.7	15
71	Amyotrophic Lateral Sclerosis: Pathogenesis, Differential Diagnoses, And Potential Interventions. <i>Journal of Spinal Cord Medicine</i> , 2002, 25, 262-273.	1.4	12
72	Serial in vivo imaging of transplanted allogeneic neural stem cell survival in a mouse model of amyotrophic lateral sclerosis. <i>Experimental Neurology</i> , 2017, 289, 96-102.	4.1	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.	3.3	11
74	Edaravone activates the GDNF/RET neurotrophic signaling pathway and protects mRNA-induced motor neurons from iPS cells. Molecular Neurodegeneration, 2022, 17, 8.	10.8	10
75	Focal and dose-dependent neuroprotection in ALS mice following AAV2-neurturin delivery. Experimental Neurology, 2020, 323, 113091.	4.1	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.	10.2	9
77	Hemiparetic Primary Lateral Sclerosis: Revisiting Mills Syndrome. Case Reports in Neurology, 2015, 7, 191-195.	0.7	7
78	Stem Cell Transplantation for Spinal Cord Neurodegeneration. Methods in Molecular Biology, 2011, 793, 479-493.	0.9	6
79	Exploring Motor Neuron Diseases Using iPSC Platforms. Stem Cells, 2022, 40, 2-13.	3.2	6
80	Motoneuron Disease: Basic Science. Advances in Neurobiology, 2017, 15, 163-190.	1.8	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, , .	0.3	5
83	Motoneuron Disease: Clinical. Advances in Neurobiology, 2017, 15, 191-210.	1.8	2
84	Rethinking a drug treatment failure on a traditional ALS target. Experimental Neurology, 2009, 216, 254-257.	4.1	1
85	New Treatments in Amyotrophic Lateral Sclerosis. Neuropsychopharmacology, 2011, 36, 370-372.	5.4	1
86	Erratum. Advances in Neurobiology, 2017, 15, E1-E1.	1.8	1
87	Human induced pluripotent stem cellâ€‘derived astrocytes progenitors as discovery platforms. , 2022, , 45-89.		0