Ernesto Bongarzone

List of Publications by Year
in descending order

57	2,073	230014	274796
papers	citations	27 h-index	44 g-index

58	58	58	2572
all docs	docs citations	times ranked	citing authors

#	Article	IF	Citations
1	The Pathogenic Sphingolipid Psychosine is Secreted in Extracellular Vesicles in the Brain of a Mouse Model of Krabbe Disease. ASN Neuro, 2022, 14, 175909142210878.	1.5	7
2	CRISPR-Cas9 Knock-In of T513M and G41S Mutations in the Murine Î²â€“Galactosyl-Ceramidase Gene Re-capitulates Early-Onset and Adult-Onset Forms of Krabbe Disease. Frontiers in Molecular Neuroscience, 2022, 15, .	1.4	5
3	Synaptic Function and Dysfunction in Lysosomal Storage Diseases. Frontiers in Cellular Neuroscience, 2021, 15, 619777.	1.8	9
4	Waning efficacy in a long-term AAV-mediated gene therapy study in the murine model of Krabbe disease. Molecular Therapy, 2021, 29, 1883-1902.	3.7	22
5	Krabbe disease: New hope for an old disease. Neuroscience Letters, 2021, 752, 135841.	1.0	34
6	Biochemical Analysis of Lipid Rafts to Study Pathogenic Mechanisms of Neural Diseases. Methods in Molecular Biology, 2021, 2187, 37-46.	0.4	2
7	The Role of Vesicle Trafficking and Release in Oligodendrocyte Biology. Neurochemical Research, 2020, 45, 620-629.	1.6	15
8	Deregulation of signalling in genetic conditions affecting the lysosomal metabolism of cholesterol and galactosyl-sphingolipids. Neurobiology of Disease, 2020, 146, 105142.	2.1	6
9	Brainstem development requires galactosylceramidase and is critical for pathogenesis in a model of Krabbe disease. Nature Communications, 2020, 11, 5356.	5.8	21
10	AAV-Mediated GALC Gene Therapy Rescues Alpha-Synucleinopathy in the Spinal Cord of a Leukodystrophic Lysosomal Storage Disease Mouse Model. Frontiers in Cellular Neuroscience, 2020, 14, 619712.	1.8	5
11	Mass spectrometry imaging and LC/MS reveal decreased cerebellar phosphoinositides in Niemann-Pick type C1-null mice. Journal of Lipid Research, 2020, 61, 1004-1013.	2.0	7
12	Lead Optimization of Benzoxazolone Carboxamides as Orally Bioavailable and CNS Penetrant Acid Ceramidase Inhibitors. Journal of Medicinal Chemistry, 2020, 63, 3634-3664.	2.9	11
13	Astrocyte Support for Oligodendrocyte Differentiation can be Conveyed via Extracellular Vesicles but Diminishes with Age. Scientific Reports, 2020, 10, 828.	1.6	53
14	Macrophages Expressing GALC Improve Peripheral Krabbe Disease by a Mechanism Independent of Cross-Correction. Neuron, 2020, 107, 65-81.e9.	3.8	39
15	Krabbe disease successfully treated via monotherapy of intrathecal gene therapy. Journal of Clinical Investigation, 2020, 130, 4906-4920.	3.9	41
16	Standard-flow LC and thermal focusing ESI elucidates altered liver proteins in late stage Niemannâ€“Pick, type C1 disease. Bioanalysis, 2019, 11, 1067-1083.	0.6	13
17	Extracellular vesicle fibrinogen induces encephalitogenic CD8+ T cells in a mouse model of multiple sclerosis. Proceedings of the National Academy of Sciences of the United States of America, 2019, 116, 10488-10493.	3.3	54
18	Inhibition of IGF-1-PI3K-Akt-mTORC2 in lipid rafts increases neuronal vulnerability in a genetic lysosomal glycosphingolipidosis. DMM Disease Models and Mechanisms, 2019, 12, .	1.2	26

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19	Long-Term Improvement of Neurological Signs and Metabolic Dysfunction in a Mouse Model of Krabbeâ€™s Disease after Global Gene Therapy. Molecular Therapy, 2018, 26, 874-889.	3.7	50
20	microRNA-219 Reduces Viral Load and Pathologic Changes in Theiler's Virus-Induced Demyelinating Disease. Molecular Therapy, 2018, 26, 730-743.	3.7	13
21	SVCT2 Expression and Function in Reactive Astrocytes Is a Common Event in Different Brain Pathologies. Molecular Neurobiology, 2018, 55, 5439-5452.	1.9	20
22	AAVrh10 Gene Therapy Ameliorates Central and Peripheral Nervous System Disease in Canine Globoid Cell Leukodystrophy (Krabbe Disease). Human Gene Therapy, 2018, 29, 785-801.	1.4	56
23	Psychosine remodels model lipid membranes at neutral pH. Biochimica Et Biophysica Acta - Biomembranes, 2018, 1860, 2515-2526.	1.4	9
24	Î±-Synuclein interacts directly but reversibly with psychosine: implications for Î±-synucleinopathies. Scientific Reports, 2018, 8, 12462.	1.6	28
25	Analysis of age-related changes in psychosine metabolism in the human brain. PLoS ONE, 2018, 13, e0193438.	1.1	24
26	miR-219 Cooperates with miR-338 in Myelination and Promotes Myelin Repair in the CNS. Developmental Cell, 2017, 40, 566-582.e5.	3.1	129
27	Psychosine enhances the shedding of membrane microvesicles: Implications in demyelination in Krabbeâ€™s disease. PLoS ONE, 2017, 12, e0178103.	1.1	28
28	Fluid levity of the cell: Role of membrane lipid architecture in genetic sphingolipidoses. Journal of Neuroscience Research, 2016, 94, 1019-1024.	1.3	11
29	How membrane dysfunction influences neuronal survival pathways in sphingolipid storage disorders. Journal of Neuroscience Research, 2016, 94, 1042-1048.	1.3	14
30	Generation of a LacZ reporter transgenic mouse line for the stereological analysis of oligodendrocyte loss in galactosylceramidase deficiency. Journal of Neuroscience Research, 2016, 94, 1520-1530.	1.3	4
31	Sulfatides in extracellular vesicles isolated from plasma of multiple sclerosis patients. Journal of Neuroscience Research, 2016, 94, 1579-1587.	1.3	45
32	Hematopoietic Stem cell transplantation and lentiviral vectorâ€based gene therapy for Krabbe's disease: Present convictions and future prospects. Journal of Neuroscience Research, 2016, 94, 1152-1168.	1.3	18
33	Intrathecal administration of AAV/GALC vectors in 10â€“11â€dayâ€old twitcher mice improves survival and is enhanced by bone marrow transplant. Journal of Neuroscience Research, 2016, 94, 1138-1151.	1.3	42
34	Beyond Krabbe's disease: The potential contribution of galactosylceramidase deficiency to neuronal vulnerability in lateâ€onset synucleinopathies. Journal of Neuroscience Research, 2016, 94, 1328-1332.	1.3	39
35	Synaptic failure: The achilles tendon of sphingolipidoses. Journal of Neuroscience Research, 2016, 94, 1031-1036.	1.3	12
36	A tribute to the work and life of Dr. Knud H. Krabbe: Advances in genetics, neuropathogenesis, therapies, and clinical management of Krabbe's disease. Journal of Neuroscience Research, 2016, 94, 963-964.	1.3	1

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37	SVCT2 Overexpression in Neuroblastoma Cells Induces Cellular Branching that is Associated with ERK Signaling. Molecular Neurobiology, 2016, 53, 6668-6679.	1.9	15
38	Recent progress on lipid lateral heterogeneity in plasma membranes: From rafts to submicrometric domains. Progress in Lipid Research, 2016, 62, 1-24.	5.3	134
39	A microglial hypothesis of globoid cell leukodystrophy pathology. Journal of Neuroscience Research, 2016, 94, 1049-1061.	1.3	24
40	Mechanism of Neuromuscular Dysfunction in Krabbe Disease. Journal of Neuroscience, 2015, 35, 1606-1616.	1.7	30
41	Unconventional Neurogenic Niches and Neurogenesis Modulation by Vitamins. Journal of Stem Cell Research & Therapy, 2014, 04, 184.	0.3	17
42	Aberrant Production of Tenascin-C in Globoid Cell Leukodystrophy Alters Psychosine-Induced Microglial Functions. Journal of Neuropathology and Experimental Neurology, 2014, 73, 964-974.	0.9	30
43	Vitamin C Transporters, Recycling and the Bystander Effect in the Nervous System: SVCT2 versus Gluts. Journal of Stem Cell Research & Therapy, 2014, 04, 209.	0.3	67
44	Neuronal inclusions of Î±â€synuclein contribute to the pathogenesis of Krabbe disease. Journal of Pathology, 2014, 232, 509-521.	2.1	89
45	Characterization and application of a disease-cell model for a neurodegenerative lysosomal disease. Molecular Genetics and Metabolism, 2014, 111, 172-183.	0.5	29
46	An In Vitro Model for the Study of Cellular Pathophysiology in Globoid Cell Leukodystrophy. Journal of Visualized Experiments, 2014, , e51903.	0.2	2
47	The Sphingolipid Psychosine Inhibits Fast Axonal Transport in Krabbe Disease by Activation of GSK3Â and Deregulation of Molecular Motors. Journal of Neuroscience, 2013, 33, 10048-10056.	1.7	80
48	Psychosine, the cytotoxic sphingolipid that accumulates in globoid cell leukodystrophy, alters membrane architecture. Journal of Lipid Research, 2013, 54, 3303-3311.	2.0	61
49	MMPâ€3 mediates psychosineâ€induced globoid cell formation: Implications for leukodystrophy pathology. Glia, 2013, 61, 765-777.	2.5	33
50	Detection of the Neurotoxin Psychosine in Samples of Peripheral Blood: Application in Diagnostics and Follow-up of Krabbe Disease. Archives of Pathology and Laboratory Medicine, 2012, 136, 709-710.	1.2	19
51	Psychosine induces the dephosphorylation of neurofilaments by deregulation of PP1 and PP2A phosphatases. Neurobiology of Disease, 2012, 46, 325-335.	2.1	44
52	Persistence of psychosine in brain lipid rafts is a limiting factor in the therapeutic recovery of a mouse model for Krabbe disease. Journal of Neuroscience Research, 2011, 89, 352-364.	1.3	54
53	Peripheral Neuropathy in the Twitcher Mouse Involves the Activation of Axonal Caspase 3. ASN Neuro, 2011, 3, AN20110019.	1.5	48
54	Psychosine Accumulates in Membrane Microdomains in the Brain of Krabbe Patients, Disrupting the Raft Architecture. Journal of Neuroscience, 2009, 29, 6068-6077.	1.7	140

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55	Combined hematopoietic and lentiviral geneâ€transfer therapies in newborn Twitcher mice reveal contemporaneous neurodegeneration and demyelination in Krabbe disease. Journal of Neuroscience Research, 2009, 87, 1748-1759.	1.3	72
56	Autonomic Denervation of Lymphoid Organs Leads to Epigenetic Immune Atrophy in a Mouse Model of Krabbe Disease. Journal of Neuroscience, 2007, 27, 13730-13738.	1.7	51
57	Central nervous system myelination in mice with deficient expression of Notch1 receptor. Journal of Neuroscience Research, 2002, 67, 309-320.	1.3	121