

# Moacir Wajner

## List of Publications by Year in descending order

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397  
papers

10,674  
citations

38660

50  
h-index

91712

69  
g-index

398  
all docs

398  
docs citations

398  
times ranked

7978  
citing authors

#	ARTICLE	IF	CITATIONS
1	Natural History, Outcome, and Treatment Efficacy in Children and Adults with Glutaryl-CoA Dehydrogenase Deficiency. <i>Pediatric Research</i> , 2006, 59, 840-847.	1.1	224
2	l-carnitine supplementation as a potential antioxidant therapy for inherited neurometabolic disorders. <i>Gene</i> , 2014, 533, 469-476.	1.0	180
3	Mitochondrial dysfunction in fatty acid oxidation disorders: insights from human and animal studies. <i>Bioscience Reports</i> , 2016, 36, e00281.	1.1	138
4	Methylmalonate administration decreases Na <sup>+</sup> ,K <sup>+</sup> -ATPase activity in cerebral cortex of rats. <i>NeuroReport</i> , 2000, 11, 2331-2334.	0.6	119
5	Resveratrol Protects C6 Astrocyte Cell Line against Hydrogen Peroxide-Induced Oxidative Stress through Heme Oxygenase 1. <i>PLoS ONE</i> , 2013, 8, e64372.	1.1	114
6	Guanosine enhances glutamate uptake in brain cortical slices at normal and excitotoxic conditions. <i>Cellular and Molecular Neurobiology</i> , 2002, 22, 353-363.	1.7	109
7	An overview of L-2-hydroxyglutarate dehydrogenase gene (L2HGDH) variants: a genotype-phenotype study. <i>Human Mutation</i> , 2010, 31, 380-390.	1.1	108
8	Inhibition of Na <sup>(+)</sup> ,K <sup>(+)</sup> -ATPase activity in hippocampus of rats subjected to acute administration of homocysteine is prevented by vitamins E and C treatment. <i>Neurochemical Research</i> , 2002, 27, 1685-1689.	1.6	96
9	D-2-hydroxyglutaric acid induces oxidative stress in cerebral cortex of young rats. <i>European Journal of Neuroscience</i> , 2003, 17, 2017-2022.	1.2	95
10	Distribution of xanthine dehydrogenase and oxidase activities in human and rabbit tissues. <i>Biochimica Et Biophysica Acta - General Subjects</i> , 1989, 991, 79-84.	1.1	93
11	<scp>l</scp>-2-Hydroxyglutaric Aciduria: Pattern of MR Imaging Abnormalities in 56 Patients. <i>Radiology</i> , 2009, 251, 856-865.	3.6	90
12	Quinolinic acid inhibits glutamate uptake into synaptic vesicles from rat brain. <i>NeuroReport</i> , 2000, 11, 249-254.	0.6	86
13	In vitro effect of homocysteine on some parameters of oxidative stress in rat hippocampus. <i>Metabolic Brain Disease</i> , 2003, 18, 147-154.	1.4	84
14	Mitochondrial energy metabolism is markedly impaired by d-2-hydroxyglutaric acid in rat tissues. <i>Molecular Genetics and Metabolism</i> , 2005, 86, 188-199.	0.5	84
15	Propionic and L-methylmalonic acids induce oxidative stress in brain of young rats. <i>NeuroReport</i> , 2000, 11, 541-544.	0.6	82
16	Reduction of Na <sup>(+)</sup> ,K <sup>(+)</sup> -ATPase activity in hippocampus of rats subjected to chemically induced hyperhomocysteinemia. <i>Neurochemical Research</i> , 2002, 27, 1593-1598.	1.6	82
17	Glutaric acid induces oxidative stress in brain of young rats. <i>Brain Research</i> , 2003, 964, 153-158.	1.1	79
18	Inhibition of brain energy metabolism by the $\hat{\alpha}$ -keto acids accumulating in maple syrup urine disease. <i>Biochimica Et Biophysica Acta - Molecular Basis of Disease</i> , 2003, 1639, 232-238.	1.8	79

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19	Mitochondrial permeability transition in neuronal damage promoted by Ca <sup>2+</sup> and respiratory chain complex II inhibition. <i>Journal of Neurochemistry</i> , 2004, 90, 1025-1035.	2.1	79
20	Inhibition of cytochrome c oxidase activity in rat cerebral cortex and human skeletal muscle by d-2-hydroxyglutaric acid in vitro. <i>Biochimica Et Biophysica Acta - Molecular Basis of Disease</i> , 2002, 1586, 81-91.	1.8	77
21	In vitro evidence for an antioxidant role of 3-hydroxykynurenine and 3-hydroxyanthranilic acid in the brain. <i>Neurochemistry International</i> , 2007, 50, 83-94.	1.9	77
22	Evidence that oxidative stress is increased in plasma from patients with maple syrup urine disease. <i>Metabolic Brain Disease</i> , 2006, 21, 279-286.	1.4	75
23	Î±-Ketoisocaproic acid and leucine provoke mitochondrial bioenergetic dysfunction in rat brain. <i>Brain Research</i> , 2010, 1324, 75-84.	1.1	75
24	Induction of oxidative stress in rat brain by the metabolites accumulating in maple syrup urine disease. <i>International Journal of Developmental Neuroscience</i> , 2003, 21, 327-332.	0.7	73
25	Disruption of mitochondrial homeostasis in organic acidurias: insights from human and animal studies. <i>Journal of Bioenergetics and Biomembranes</i> , 2011, 43, 31-38.	1.0	71
26	Intrastriatal methylmalonic acid administration induces rotational behavior and convulsions through glutamatergic mechanisms. <i>Brain Research</i> , 1996, 721, 120-125.	1.1	69
27	Î±-Keto Acids Accumulating in Maple Syrup Urine Disease Stimulate Lipid Peroxidation and Reduce Antioxidant Defences in Cerebral Cortex From Young Rats. <i>Metabolic Brain Disease</i> , 2005, 20, 155-167.	1.4	69
28	Oxidative Stress in Phenylketonuria: What is the Evidence?. <i>Cellular and Molecular Neurobiology</i> , 2011, 31, 653-662.	1.7	67
29	3-hydroxyglutaric acid induces oxidative stress and decreases the antioxidant defenses in cerebral cortex of young rats. <i>Brain Research</i> , 2002, 956, 367-373.	1.1	63
30	Stimulation of lipid peroxidation in vitro in rat brain by the metabolites accumulating in maple syrup urine disease. <i>Metabolic Brain Disease</i> , 2002, 17, 47-54.	1.4	63
31	Mitochondrial energy metabolism in neurodegeneration associated with methylmalonic acidemia. <i>Journal of Bioenergetics and Biomembranes</i> , 2011, 43, 39-46.	1.0	62
32	Evidence that folic acid deficiency is a major determinant of hyperhomocysteinemia in Parkinson's disease. <i>Metabolic Brain Disease</i> , 2009, 24, 257-269.	1.4	61
33	Inhibition of glutamate uptake into synaptic vesicles of rat brain by the metabolites accumulating in maple syrup urine disease. <i>Journal of the Neurological Sciences</i> , 2000, 181, 44-49.	0.3	60
34	Ascorbic acid prevents cognitive deficits caused by chronic administration of propionic acid to rats in the water maze. <i>Pharmacology Biochemistry and Behavior</i> , 2002, 73, 623-629.	1.3	60
35	Acute intrastriatal administration of quinolinic acid provokes hyperphosphorylation of cytoskeletal intermediate filament proteins in astrocytes and neurons of rats. <i>Experimental Neurology</i> , 2010, 224, 188-196.	2.0	60
36	Ascorbic acid and Î±-tocopherol attenuate methylmalonic acid-induced convulsions. <i>NeuroReport</i> , 1999, 10, 2039-2043.	0.6	59

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37	l-Carnitine Blood Levels and Oxidative Stress in Treated Phenylketonuric Patients. Cellular and Molecular Neurobiology, 2009, 29, 211-218.	1.7	59
38	Experimental hyperphenylalaninemia provokes oxidative stress in rat brain. Biochimica Et Biophysica Acta - Molecular Basis of Disease, 2002, 1586, 344-352.	1.8	58
39	Experimental Evidence that Phenylalanine Provokes Oxidative Stress in Hippocampus and Cerebral Cortex of Developing Rats. Cellular and Molecular Neurobiology, 2010, 30, 317-326.	1.7	58
40	Effect of Chemically Induced Propionic Acidemia on Neurobehavioral Development of Rats. Pharmacology Biochemistry and Behavior, 1999, 64, 529-534.	1.3	56
41	Inhibition of Brain Energy Metabolism by the Branched-chain Amino Acids Accumulating in Maple Syrup Urine Disease. Neurochemical Research, 2008, 33, 114-124.	1.6	56
42	Induction of oxidative stress by L-2-hydroxyglutaric acid in rat brain. Journal of Neuroscience Research, 2003, 74, 103-110.	1.3	55
43	Evidence that quinolinic acid severely impairs energy metabolism through activation of NMDA receptors in striatum from developing rats. Journal of Neurochemistry, 2006, 99, 1531-1542.	2.1	55
44	Chronic treatment with glutaric acid induces partial tolerance to excitotoxicity in neuronal cultures from chick embryo telencephalons. Journal of Neuroscience Research, 2002, 68, 424-431.	1.3	53
45	Proline induces oxidative stress in cerebral cortex of rats. International Journal of Developmental Neuroscience, 2003, 21, 105-110.	0.7	53
46	Inhibition of Na <sup>+</sup> ,K <sup>+</sup> -ATPase from rat brain cortex by propionic acid. NeuroReport, 1998, 9, 1719-1721.	0.6	52
47	Inhibition of synaptosomal [3H]glutamate uptake and [3H]glutamate binding to plasma membranes from brain of young rats by glutaric acid in vitro. Journal of the Neurological Sciences, 2000, 173, 93-96.	0.3	52
48	Evaluation of the mechanisms involved in leucine-induced oxidative damage in cerebral cortex of young rats. Free Radical Research, 2005, 39, 71-79.	1.5	52
49	Effect of short and long term exposition to high phenylalanine blood levels on oxidative damage in phenylketonuric patients. International Journal of Developmental Neuroscience, 2009, 27, 243-247.	0.7	52
50	Induction of Oxidative Stress by Chronic and Acute Glutaric Acid Administration to Rats. Cellular and Molecular Neurobiology, 2007, 27, 423-438.	1.7	51
51	Inhibition of Na <sup>+</sup> , K <sup>+</sup> -ATPase activity by the metabolites accumulating in homocystinuria. Metabolic Brain Disease, 2002, 17, 83-91.	1.4	49
52	Prevention by l-carnitine of DNA damage induced by propionic and l-methylmalonic acids in human peripheral leukocytes in vitro. Mutation Research - Genetic Toxicology and Environmental Mutagenesis, 2010, 702, 123-128.	0.9	49
53	Experimental Evidence that Methylmalonic Acid Provokes Oxidative Damage and Compromises Antioxidant Defenses in Nerve Terminal and Striatum of Young Rats. Cellular and Molecular Neurobiology, 2011, 31, 775-785.	1.7	49
54	Neurological Damage in MSUD: The Role of Oxidative Stress. Cellular and Molecular Neurobiology, 2014, 34, 157-165.	1.7	49

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55	Morphological alterations and induction of oxidative stress in glial cells caused by the branched-chain $\alpha$ -keto acids accumulating in maple syrup urine disease. <i>Neurochemistry International</i> , 2006, 49, 640-650.	1.9	48
56	Guanidinoacetate Decreases Antioxidant Defenses and Total Protein Sulfhydryl Content in Striatum of Rats. <i>Neurochemical Research</i> , 2008, 33, 1804-1810.	1.6	48
57	Cannabinoid receptor agonists reduce the short-term mitochondrial dysfunction and oxidative stress linked to excitotoxicity in the rat brain. <i>Neuroscience</i> , 2015, 285, 97-106.	1.1	48
58	Pharmacological evidence for GABAergic and glutamatergic involvement in the convulsant and behavioral effects of glutaric acid. <i>Brain Research</i> , 1998, 802, 55-60.	1.1	47
59	Differential inhibitory effects of methylmalonic acid on respiratory chain complex activities in rat tissues. <i>International Journal of Developmental Neuroscience</i> , 2006, 24, 45-52.	0.7	47
60	Selective screening for organic acidemias by urine organic acid GC-MS analysis in Brazil: Fifteen-year experience. <i>Clinica Chimica Acta</i> , 2009, 400, 77-81.	0.5	47
61	Inhibition of rat brain Na <sup>+</sup> , K <sup>+</sup> -ATPase activity induced by homocysteine is probably mediated by oxidative stress. <i>Neurochemical Research</i> , 2001, 26, 1195-1200.	1.6	46
62	Brain energy metabolism is compromised by the metabolites accumulating in homocystinuria. <i>Neurochemistry International</i> , 2003, 43, 597-602.	1.9	45
63	Quinolinic acid reduces the antioxidant defenses in cerebral cortex of young rats. <i>International Journal of Developmental Neuroscience</i> , 2005, 23, 695-701.	0.7	45
64	Astrocytic proliferation and mitochondrial dysfunction induced by accumulated glutaric acidemia I (GAI) metabolites: Possible implications for GAI pathogenesis. <i>Neurobiology of Disease</i> , 2008, 32, 528-534.	2.1	45
65	Reduction of lipid and protein damage in patients with disorders of propionate metabolism under treatment: a possible protective role of l-carnitine supplementation. <i>International Journal of Developmental Neuroscience</i> , 2010, 28, 127-132.	0.7	45
66	Methionine alters Na <sup>+</sup> , K <sup>+</sup> -ATPase activity, lipid peroxidation and nonenzymatic antioxidant defenses in rat hippocampus. <i>International Journal of Developmental Neuroscience</i> , 2005, 23, 651-656.	0.7	44
67	In vitro phosphorylation of cytoskeletal proteins from cerebral cortex of rats. <i>Brain Research Protocols</i> , 2003, 11, 111-118.	1.7	43
68	Neurological manifestations of organic acidurias. <i>Nature Reviews Neurology</i> , 2019, 15, 253-271.	4.9	43
69	Effect of proline administration on rat behavior in aversive and nonaversive tasks. <i>Pharmacology Biochemistry and Behavior</i> , 1989, 32, 885-890.	1.3	42
70	Arginine Administration Decreases Cerebral Cortex Acetylcholinesterase and Serum Butyrylcholinesterase Probably by Oxidative Stress Induction. <i>Neurochemical Research</i> , 2004, 29, 385-389.	1.6	42
71	Evidence that 3-hydroxyglutaric acid interacts with NMDA receptors in synaptic plasma membranes from cerebral cortex of young rats. <i>Neurochemistry International</i> , 2004, 45, 1087-1094.	1.9	42
72	Inhibition of creatine kinase activity from rat cerebral cortex by $\alpha$ -2-hydroxyglutaric acid in vitro. <i>Neurochemistry International</i> , 2004, 44, 45-52.	1.9	42

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73	$\beta^3$ -Hydroxybutyric acid induces oxidative stress in cerebral cortex of young rats. <i>Neurochemistry International</i> , 2007, 50, 564-570.	1.9	42
74	Lipoic acid prevents oxidative stress in vitro and in vivo by an acute hyperphenylalaninemia chemically-induced in rat brain. <i>Journal of the Neurological Sciences</i> , 2010, 292, 89-95.	0.3	42
75	Urinary biomarkers of oxidative stress and plasmatic inflammatory profile in phenylketonuric treated patients. <i>International Journal of Developmental Neuroscience</i> , 2015, 47, 259-265.	0.7	42
76	Bezafibrate prevents mitochondrial dysfunction, antioxidant system disturbance, glial reactivity and neuronal damage induced by sulfite administration in striatum of rats: Implications for a possible therapeutic strategy for sulfite oxidase deficiency. <i>Biochimica Et Biophysica Acta - Molecular Basis of Disease</i> , 2017, 1863, 2135-2148.	1.8	42
77	Nitric oxide synthase inhibition by L-NAME prevents the decrease of Na <sup>+</sup> ,K <sup>+</sup> -ATPase activity in midbrain of rats subjected to arginine administration. <i>Neurochemical Research</i> , 2001, 26, 515-520.	1.6	41
78	Intrastratial administration of 3-hydroxyglutaric acid induces convulsions and striatal lesions in rats. <i>Brain Research</i> , 2001, 916, 70-75.	1.1	41
79	Antioxidant Effect of Cysteamine in Brain Cortex of Young Rats. <i>Neurochemical Research</i> , 2008, 33, 737-744.	1.6	41
80	Evidence that DNA damage is associated to phenylalanine blood levels in leukocytes from phenylketonuric patients. <i>Mutation Research - Genetic Toxicology and Environmental Mutagenesis</i> , 2009, 679, 13-16.	0.9	41
81	Inhibition of creatine kinase activity in vitro by ethylmalonic acid in cerebral cortex of young rats. <i>Neurochemical Research</i> , 2002, 27, 1633-1639.	1.6	40
82	Protein and lipid damage in maple syrup urine disease patients: carnitine effect. <i>International Journal of Developmental Neuroscience</i> , 2013, 31, 21-24.	0.7	40
83	In vitro inhibition of Na <sup>+</sup> ,K <sup>+</sup> -ATPase activity from rat cerebral cortex by guanidino compounds accumulating in hyperargininemia. <i>Brain Research</i> , 1999, 838, 78-84.	1.1	39
84	Isovaleric Acid Reduces Na <sup>+</sup> , K <sup>+</sup> -ATPase Activity in Synaptic Membranes from Cerebral Cortex of Young Rats. <i>Cellular and Molecular Neurobiology</i> , 2007, 27, 529-540.	1.7	39
85	Oxidative stress in plasma from maple syrup urine disease patients during treatment. <i>Metabolic Brain Disease</i> , 2008, 23, 71-80.	1.4	39
86	Long-chain 3-hydroxy fatty acids accumulating in long-chain 3-hydroxyacyl-CoA dehydrogenase and mitochondrial trifunctional protein deficiencies uncouple oxidative phosphorylation in heart mitochondria. <i>Journal of Bioenergetics and Biomembranes</i> , 2013, 45, 47-57.	1.0	39
87	Impairment of energy metabolism in hippocampus of rats subjected to chemically-induced hyperhomocysteinemia. <i>Biochimica Et Biophysica Acta - Molecular Basis of Disease</i> , 2003, 1637, 187-192.	1.8	38
88	Experimental evidence of oxidative stress in plasma of homocystinuric patients: A possible role for homocysteine. <i>Molecular Genetics and Metabolism</i> , 2011, 104, 112-117.	0.5	38
89	Oxidative stress in Niemann-Pick type C patients: a protective role of N-butyldeoxynojirimycin therapy. <i>International Journal of Developmental Neuroscience</i> , 2012, 30, 439-444.	0.7	38
90	Inhibition of the mitochondrial respiratory chain by phenylalanine in rat cerebral cortex. <i>Neurochemical Research</i> , 2002, 27, 353-357.	1.6	37

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91	Intrastriatal Administration of Guanidinoacetate Inhibits Na <sup>+</sup> , K <sup>+</sup> -ATPase and Creatine Kinase Activities in Rat Striatum. <i>Metabolic Brain Disease</i> , 2006, 21, 39-48.	1.4	37
92	Proline reduces acetylcholinesterase activity in cerebral cortex of rats. <i>Metabolic Brain Disease</i> , 2003, 18, 79-86.	1.4	36
93	Evidence for a synergistic action of glutaric and 3- $\alpha$ -hydroxyglutaric acids disturbing rat brain energy metabolism. <i>International Journal of Developmental Neuroscience</i> , 2007, 25, 391-398.	0.7	36
94	Creatine kinase activity from rat brain is inhibited by branched-chain amino acids in vitro. <i>Neurochemical Research</i> , 2003, 28, 675-679.	1.6	35
95	Inhibition of energy metabolism in cerebral cortex of young rats by the medium-chain fatty acids accumulating in MCAD deficiency. <i>Brain Research</i> , 2004, 1030, 141-151.	1.1	35
96	Induction of lipid peroxidation and decrease of antioxidant defenses in symptomatic and asymptomatic patients with X-linked adrenoleukodystrophy. <i>International Journal of Developmental Neuroscience</i> , 2007, 25, 441-444.	0.7	35
97	Induction of oxidative stress by the metabolites accumulating in 3-methylglutaconic aciduria in cerebral cortex of young rats. <i>Life Sciences</i> , 2008, 82, 652-662.	2.0	35
98	Disturbance of brain energy and redox homeostasis provoked by sulfite and thiosulfate: Potential pathomechanisms involved in the neuropathology of sulfite oxidase deficiency. <i>Gene</i> , 2013, 531, 191-198.	1.0	35
99	Oxidative Stress, Disrupted Energy Metabolism, and Altered Signaling Pathways in Glutaryl-CoA Dehydrogenase Knockout Mice: Potential Implications of Quinolinic Acid Toxicity in the Neuropathology of Glutaric Acidemia Type I. <i>Molecular Neurobiology</i> , 2016, 53, 6459-6475.	1.9	35
100	Characterization of the inhibition of pyruvate kinase caused by phenylalanine and phenylpyruvate in rat brain cortex. <i>Brain Research</i> , 2003, 968, 199-205.	1.1	34
101	5-Oxoproline Reduces Non-Enzymatic Antioxidant Defenses in vitro in Rat Brain. <i>Metabolic Brain Disease</i> , 2007, 22, 51-65.	1.4	34
102	Signaling mechanisms downstream of quinolinic acid targeting the cytoskeleton of rat striatal neurons and astrocytes. <i>Experimental Neurology</i> , 2012, 233, 391-399.	2.0	34
103	Signaling mechanisms underlying the glioprotective effects of resveratrol against mitochondrial dysfunction. <i>Biochimica Et Biophysica Acta - Molecular Basis of Disease</i> , 2016, 1862, 1827-1838.	1.8	34
104	In vivo and in vitro effects of proline on some parameters of oxidative stress in rat brain. <i>Brain Research</i> , 2003, 991, 180-186.	1.1	33
105	Glutaric acid stimulates glutamate binding and astrocytic uptake and inhibits vesicular glutamate uptake in forebrain from young rats. <i>Neurochemistry International</i> , 2004, 45, 1075-1086.	1.9	33
106	Evidence that the major metabolites accumulating in medium-chain acyl-CoA dehydrogenase deficiency disturb mitochondrial energy homeostasis in rat brain. <i>Brain Research</i> , 2009, 1296, 117-126.	1.1	33
107	Neurochemical evidence that phytanic acid induces oxidative damage and reduces the antioxidant defenses in cerebellum and cerebral cortex of rats. <i>Life Sciences</i> , 2010, 87, 275-280.	2.0	33
108	Chemically induced model of hypermethioninemia in rats. <i>Journal of Neuroscience Methods</i> , 2007, 160, 1-4.	1.3	32



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109	Tyrosine promotes oxidative stress in cerebral cortex of young rats. <i>International Journal of Developmental Neuroscience</i> , 2008, 26, 551-559.	0.7	32
110	Medium-chain fatty acids accumulating in MCAD deficiency elicit lipid and protein oxidative damage and decrease non-enzymatic antioxidant defenses in rat brain. <i>Neurochemistry International</i> , 2009, 54, 519-525.	1.9	32
111	Lower in vivo brain extracellular GABA concentration in diabetic rats during forced swimming. <i>Brain Research</i> , 2003, 968, 281-284.	1.1	31
112	Glutaric Acid Administration Impairs Energy Metabolism in Midbrain and Skeletal Muscle of Young Rats. <i>Neurochemical Research</i> , 2005, 30, 1123-1131.	1.6	31
113	Evidence that glutaric acid reduces glutamate uptake by cerebral cortex of infant rats. <i>Life Sciences</i> , 2007, 81, 1668-1676.	2.0	31
114	Acute administration of 5-oxoproline induces oxidative damage to lipids and proteins and impairs antioxidant defenses in cerebral cortex and cerebellum of young rats. <i>Metabolic Brain Disease</i> , 2010, 25, 145-154.	1.4	31
115	Sulfite disrupts brain mitochondrial energy homeostasis and induces mitochondrial permeability transition pore opening via thiol group modification. <i>Biochimica Et Biophysica Acta - Molecular Basis of Disease</i> , 2014, 1842, 1413-1422.	1.8	31
116	Promotion of oxidative stress by l-tryptophan in cerebral cortex of rats. <i>Neurochemistry International</i> , 2006, 49, 87-93.	1.9	30
117	Tyrosine administration decreases glutathione and stimulates lipid and protein oxidation in rat cerebral cortex. <i>Metabolic Brain Disease</i> , 2009, 24, 415-425.	1.4	30
118	Long-chain 3-hydroxy fatty acids accumulating in LCHAD and MTP deficiencies induce oxidative stress in rat brain. <i>Neurochemistry International</i> , 2010, 56, 930-936.	1.9	30
119	Disturbance of mitochondrial energy homeostasis caused by the metabolites accumulating in LCHAD and MTP deficiencies in rat brain. <i>Life Sciences</i> , 2010, 86, 825-831.	2.0	30
120	Oxidative Stress Parameters in Urine from Patients with Disorders of Propionate Metabolism: a Beneficial Effect of l-Carnitine Supplementation. <i>Cellular and Molecular Neurobiology</i> , 2012, 32, 77-82.	1.7	30
121	Oxidative damage in glutaric aciduria type I patients and the protective effects of l-carnitine treatment. <i>Journal of Cellular Biochemistry</i> , 2018, 119, 10021-10032.	1.2	30
122	l-2-Hydroxyglutaric acid inhibits mitochondrial creatine kinase activity from cerebellum of developing rats. <i>International Journal of Developmental Neuroscience</i> , 2003, 21, 217-224.	0.7	29
123	A chemically-induced acute model of maple syrup urine disease in rats for neurochemical studies. <i>Journal of Neuroscience Methods</i> , 2006, 155, 224-230.	1.3	29
124	Kynurenines Impair Energy Metabolism in Rat Cerebral Cortex. <i>Cellular and Molecular Neurobiology</i> , 2007, 27, 147-160.	1.7	29
125	Evidence that 3-hydroxy-3-methylglutaric acid promotes lipid and protein oxidative damage and reduces the nonenzymatic antioxidant defenses in rat cerebral cortex. <i>Journal of Neuroscience Research</i> , 2008, 86, 683-693.	1.3	29
126	Effects of 1,4-butanediol administration on oxidative stress in rat brain: Study of the neurotoxicity of ̢-hydroxybutyric acid in vivo. <i>Metabolic Brain Disease</i> , 2009, 24, 271-282.	1.4	29



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127	Simvastatin treatment prevents oxidative damage to DNA in whole blood leukocytes of dyslipidemic type 2 diabetic patients. <i>Cell Biochemistry and Function</i> , 2010, 28, 360-366.	1.4	29
128	Induction of oxidative stress in brain of glutaryl-CoA dehydrogenase deficient mice by acute lysine administration. <i>Molecular Genetics and Metabolism</i> , 2012, 106, 31-38.	0.5	29
129	Toxicity of octanoate and decanoate in rat peripheral tissues: evidence of bioenergetic dysfunction and oxidative damage induction in liver and skeletal muscle. <i>Molecular and Cellular Biochemistry</i> , 2012, 361, 329-335.	1.4	29
130	Disruption of brain redox homeostasis in glutaryl-CoA dehydrogenase deficient mice treated with high dietary lysine supplementation. <i>Molecular Genetics and Metabolism</i> , 2013, 108, 30-39.	0.5	29
131	Investigation of inflammatory profile in MSUD patients: benefit of L-carnitine supplementation. <i>Metabolic Brain Disease</i> , 2015, 30, 1167-1174.	1.4	29
132	L-Carnitine supplementation decreases DNA damage in treated MSUD patients. <i>Mutation Research - Fundamental and Molecular Mechanisms of Mutagenesis</i> , 2015, 775, 43-47.	0.4	29
133	Inhibition of Na <sup>+</sup> ,K <sup>+</sup> -ATPase activity from rat hippocampus by proline. <i>Neurochemical Research</i> , 2001, 26, 1321-1326.	1.6	28
134	In vitro stimulation of oxidative stress in cerebral cortex of rats by the guanidino compounds accumulating in hyperargininemia. <i>Brain Research</i> , 2001, 923, 50-57.	1.1	28
135	Î±-Ketoisocaproic acid regulates phosphorylation of intermediate filaments in postnatal rat cortical slices through ionotropic glutamatergic receptors. <i>Developmental Brain Research</i> , 2002, 139, 267-276.	2.1	28
136	Ethylmalonic acid inhibits mitochondrial creatine kinase activity from cerebral cortex of young rats in vitro. <i>Neurochemical Research</i> , 2003, 28, 771-777.	1.6	28
137	Ascorbic acid prevents water maze behavioral deficits caused by early postnatal methylmalonic acid administration in the rat. <i>Brain Research</i> , 2003, 976, 234-242.	1.1	28
138	Evidence that oxidative stress is involved in the inhibitory effect of proline on Na <sup>+</sup> ,K <sup>+</sup> -ATPase activity in synaptic plasma membrane of rat hippocampus. <i>International Journal of Developmental Neuroscience</i> , 2003, 21, 303-307.	0.7	28
139	Oxidative Stress in Homocystinuria Due to Cystathionine Î³-Synthase Deficiency: Findings in Patients and in Animal Models. <i>Cellular and Molecular Neurobiology</i> , 2017, 37, 1477-1485.	1.7	28
140	Neurochemical evidence that 3Î±-methylglutaric acid inhibits synaptic Na <sup>+</sup> ,K <sup>+</sup> -ATPase activity probably through oxidative damage in brain cortex of young rats. <i>International Journal of Developmental Neuroscience</i> , 2011, 29, 1-7.	0.7	27
141	Urinary biomarkers of oxidative damage in Maple syrup urine disease: The L-carnitine role. <i>International Journal of Developmental Neuroscience</i> , 2015, 42, 10-14.	0.7	27
142	2Î±-Methylcitric acid impairs glutamate metabolism and induces permeability transition in brain mitochondria. <i>Journal of Neurochemistry</i> , 2016, 137, 62-75.	2.1	27
143	Methylmalonic and propionic acids increase the in vitro incorporation of into cytoskeletal proteins from cerebral cortex of young rats through NMDA glutamate receptors. <i>Brain Research</i> , 2000, 856, 111-118.	1.1	26
144	Reduction of energy metabolism in rat hippocampus by arginine administration. <i>Brain Research</i> , 2003, 983, 58-63.	1.1	26

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146	Evidence that the branched-chain $\alpha$ -keto acids accumulating in maple syrup urine disease induce morphological alterations and death in cultured astrocytes from rat cerebral cortex. <i>Glia</i> , 2004, 48, 230-240.	2.5	26
147	Reduction of Butyrylcholinesterase Activity in Rat Serum Subjected to Hyperhomocysteinemia. <i>Metabolic Brain Disease</i> , 2005, 20, 97-103.	1.4	26
148	Amino acids levels and lipid peroxidation in maple syrup urine disease patients. <i>Clinical Biochemistry</i> , 2009, 42, 462-466.	0.8	26
149	Experimental evidence of oxidative stress in patients with l-2-hydroxyglutaric aciduria and that l-carnitine attenuates in vitro DNA damage caused by d-2-hydroxyglutaric and l-2-hydroxyglutaric acids. <i>Toxicology in Vitro</i> , 2017, 42, 47-53.	1.1	26
150	Glutaric acid moderately compromises energy metabolism in rat brain. <i>International Journal of Developmental Neuroscience</i> , 2005, 23, 687-693.	0.7	25
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157	In vitro effect of quinolinic acid on energy metabolism in brain of young rats. <i>Neuroscience Research</i> , 2007, 57, 277-288.	1.0	24
158	Glycine Provokes Lipid Oxidative Damage and Reduces the Antioxidant Defenses in Brain Cortex of Young Rats. <i>Cellular and Molecular Neurobiology</i> , 2009, 29, 253-261.	1.7	24
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248	Effects of cysteamine on oxidative status in cerebral cortex of rats. <i>Metabolic Brain Disease</i> , 2008, 23, 81-93.	1.4	15
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272	Induction of S100B secretion in C6 astroglial cells by the major metabolites accumulating in glutaric acidemia type I. <i>Metabolic Brain Disease</i> , 2010, 25, 191-198.	1.4	13
273	Phytanic acid disturbs mitochondrial homeostasis in heart of young rats: a possible pathomechanism of cardiomyopathy in Refsum disease. <i>Molecular and Cellular Biochemistry</i> , 2012, 366, 335-343.	1.4	13
274	Neurochemical Evidence that the Metabolites Accumulating in 3-Methylcrotonyl-CoA Carboxylase Deficiency Induce Oxidative Damage in Cerebral Cortex of Young Rats. <i>Cellular and Molecular Neurobiology</i> , 2013, 33, 137-146.	1.7	13
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278	Detection of Organic Acidemias in Brazil. <i>Archives of Medical Research</i> , 2002, 33, 581-585.	1.5	12
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284	Glycine Intracerebroventricular Administration Disrupts Mitochondrial Energy Homeostasis in Cerebral Cortex and Striatum of Young Rats. <i>Neurotoxicity Research</i> , 2013, 24, 502-511.	1.3	12
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288	Bezafibrate In Vivo Administration Prevents 3-Methylglutaric Acid-Induced Impairment of Redox Status, Mitochondrial Biogenesis, and Neural Injury in Brain of Developing Rats. <i>Neurotoxicity Research</i> , 2019, 35, 809-822.	1.3	12

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308	Kinetic studies on the inhibition of creatine kinase activity by branched-chain amino acids in the brain cortex of rats. <i>International Journal of Developmental Neuroscience</i> , 2003, 21, 145-151.	0.7	10
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310	Influence of ketone bodies on oxidative stress parameters in brain of developing rats in vitro. <i>Metabolic Brain Disease</i> , 2008, 23, 411-425.	1.4	10
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323	Evaluation of the effect of chronic administration of drugs on rat behavior in the water maze task. <i>Brain Research Protocols</i> , 2003, 12, 109-115.	1.7	9
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328	Evidence that glycine induces lipid peroxidation and decreases glutathione concentrations in rat cerebellum. <i>Molecular and Cellular Biochemistry</i> , 2014, 395, 125-134.	1.4	9
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331	3-Hydroxy-3-Methylglutaric Acid Impairs Redox and Energy Homeostasis, Mitochondrial Dynamics, and Endoplasmic Reticulumâ€“Mitochondria Crosstalk in Rat Brain. <i>Neurotoxicity Research</i> , 2020, 37, 314-325.	1.3	9
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339	Disturbance of mitochondrial functions associated with permeability transition pore opening induced by cis-5-tetradecenoic and myristic acids in liver of adolescent rats. <i>Mitochondrion</i> , 2020, 50, 1-13.	1.6	8
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341	Lipopolysaccharide-Elicited Systemic Inflammation Induces Selective Vulnerability of Cerebral Cortex and Striatum of Developing Glutaryl-CoA Dehydrogenase Deficient (Gcdh <sup>~</sup> / <sup>~</sup> ) Mice to Oxidative Stress. <i>Neurotoxicity Research</i> , 2020, 38, 1024-1036.	1.3	8
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363	Evaluation of Oxidative Stress Parameters and Energy Metabolism in Cerebral Cortex of Rats Subjected to Sarcosine Administration. <i>Molecular Neurobiology</i> , 2017, 54, 4496-4506.	1.9	5
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377	Inborn Errors of Metabolism. <i>Clinical Pediatrics</i> , 1989, 28, 494-497.	0.4	3
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395	Intracerebroventricular injection of glycine alters enzymatic antioxidant defenses in rat striatum: prevention by bezafibrate. <i>Free Radical Biology and Medicine</i> , 2017, 108, S34.	1.3	0
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