Bianca Seminotti

List of Publications by Year
in descending order

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216 papers	6,592 citations	57681 46 h-index	120465 65 g-index
216 all docs	216 docs citations	216 times ranked	5649 citing authors

#	Article	IF	Citations
1	Increased susceptibility to quinolinic acidâ€•induced seizures and longâ€•term changes in brain oscillations in an animal model of glutaric acidemia type I. Journal of Neuroscience Research, 2022, 100, 992-1007.	1.3	0
2	Plinia trunciflora Extract Administration Prevents HI-Induced Oxidative Stress, Inflammatory Response, Behavioral Impairments, and Tissue Damage in Rats. Nutrients, 2022, 14, 395.	1.7	2
3	Antioxidant system disturbances and mitochondrial dysfunction induced by 3-methylglutaric acid in rat heart are prevented by bezafibrate. European Journal of Pharmacology, 2022, 924, 174950.	1.7	4
4	Pathophysiology of maple syrup urine disease: Focus on the neurotoxic role of the accumulated branched-chain amino acids and branched-chain Î±-keto acids. Neurochemistry International, 2022, 157, 105360.	1.9	12
5	Clinical, biochemical and molecular findings of 24 Brazilian patients with glutaric acidemia type 1: 4 novel mutations in the GCDH gene. Metabolic Brain Disease, 2021, 36, 205-212.	1.4	7
6	The mitochondrialâ€•targeted reactive species scavenger JP4 â€039 prevents sulfiteâ€•induced alterations in antioxidant defenses, energy transfer, and cell death signaling in striatum of rats. Journal of Inherited Metabolic Disease, 2021, 44, 481-491.	1.7	7
7	Insights from Animal Models on the Pathophysiology of Hyperphenylalaninemia: Role of Mitochondrial Dysfunction, Oxidative Stress and Inflammation. Molecular Neurobiology, 2021, 58, 2897-2909.	1.9	15
8	Ethylmalonic acid impairs bioenergetics by disturbing succinate and glutamate oxidation and induces mitochondrial permeability transition pore opening in rat cerebellum. Journal of Neurochemistry, 2021, 158, 262-281.	2.1	3
9	L-carnitine protects DNA oxidative damage induced by phenylalanine and its keto acid derivatives in neural cells: a possible pathomechanism and adjuvant therapy for brain injury in phenylketonuria. Metabolic Brain Disease, 2021, 36, 1957-1968.	1.4	4
10	S-adenosylmethionine induces mitochondrial dysfunction, permeability transition pore opening and redox imbalance in subcellular preparations of rat liver. Journal of Bioenergetics and Biomembranes, 2021, 53, 525-539.	1.0	3
11	Protective effects of L-carnitine on behavioral alterations and neuroinflammation in striatum of glutaryl-COA dehydrogenase deficient mice. Archives of Biochemistry and Biophysics, 2021, 709, 108970.	1.4	5
12	Neuronal Death, Glial Reactivity, Microglia Activation, Oxidative Stress and Bioenergetics Impairment Caused by Intracerebroventricular Administration of D-2-hydroxyglutaric Acid to Neonatal Rats. Neuroscience, 2021, 471, 115-132.	1.1	8
13	Glutaric Acidemia Type 1: An Inherited Neurometabolic Disorder of Intoxication. , 2021, , 1-25.		1
14	Nuclear Factor Erythroid-2-Related Factor 2 Signaling in the Neuropathophysiology of Inherited Metabolic Disorders. Frontiers in Cellular Neuroscience, 2021, 15, 785057.	1.8	19
15	Free Radical Scavengers Prevent Argininosuccinic Acid-Induced Oxidative Stress in the Brain of Developing Rats: a New Adjuvant Therapy for Argininosuccinate Lyase Deficiency?. Molecular Neurobiology, 2020, 57, 1233-1244.	1.9	10
16	3-Hydroxy-3-Methylglutaric Acid Impairs Redox and Energy Homeostasis, Mitochondrial Dynamics, and Endoplasmic Reticulumâ€“Mitochondria Crosstalk in Rat Brain. Neurotoxicity Research, 2020, 37, 314-325.	1.3	9
17	Disturbance of mitochondrial functions associated with permeability transition pore opening induced by cis-5-tetradecenoic and myristic acids in liver of adolescent rats. Mitochondrion, 2020, 50, 1-13.	1.6	8
18	Elevated levels of BDNF and cathepsinâ€•d as possible peripheral markers of neurodegeneration in plasma of patients with glutaric acidemia type I. International Journal of Developmental Neuroscience, 2020, 80, 42-49.	0.7	8

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19	Disruption of mitochondrial functions and oxidative stress contribute to neurologic dysfunction in organic acidurias. Archives of Biochemistry and Biophysics, 2020, 696, 108646.	1.4	11
20	Mitochondrial Dysfunction and Redox Homeostasis Impairment as Pathomechanisms of Brain Damage in Ethylmalonic Encephalopathy: Insights from Animal and Human Studies. Cellular and Molecular Neurobiology, 2020, , 1.	1.7	10
21	Lipopolysaccharide-Elicited Systemic Inflammation Induces Selective Vulnerability of Cerebral Cortex and Striatum of Developing Glutaryl-CoA Dehydrogenase Deficient (Gcdhâ^'/â^') Mice to Oxidative Stress. Neurotoxicity Research, 2020, 38, 1024-1036.	1.3	8
22	Recent Advances in the Pathophysiology of Fatty Acid Oxidation Defects: Secondary Alterations of Bioenergetics and Mitochondrial Calcium Homeostasis Caused by the Accumulating Fatty Acids. Frontiers in Genetics, 2020, 11, 598976.	1.1	7
23	Guanosine Neuroprotection of Presynaptic Mitochondrial Calcium Homeostasis in a Mouse Study with Amyloid-Î² Oligomers. Molecular Neurobiology, 2020, 57, 4790-4809.	1.9	14
24	InÂvivo evidence that bezafibrate prevents oxidative stress and mitochondrial dysfunction caused by 3-methylglutaric acid in rat liver. Biochimie, 2020, 171-172, 187-196.	1.3	10
25	Disturbance of bioenergetics and calcium homeostasis provoked by metabolites accumulating in propionic acidemia in heart mitochondria of developing rats. Biochimica Et Biophysica Acta - Molecular Basis of Disease, 2020, 1866, 165682.	1.8	11
26	Disruption of Brain Redox Homeostasis, Microglia Activation and Neuronal Damage Induced by Intracerebroventricular Administration of S-Adenosylmethionine to Developing Rats. Molecular Neurobiology, 2019, 56, 2760-2773.	1.9	16
27	Long Lasting High Lysine Diet Aggravates White Matter Injury in Glutaryl-CoA Dehydrogenase Deficient (Gcdhâ^'/â^') Mice. Molecular Neurobiology, 2019, 56, 648-657.	1.9	9
28	Reticular Dysgenesis and Mitochondriopathy Induced by Adenylate Kinase 2 Deficiency with Atypical Presentation. Scientific Reports, 2019, 9, 15739.	1.6	14
29	ETHE1 and MOCS1 deficiencies: Disruption of mitochondrial bioenergetics, dynamics, redox homeostasis and endoplasmic reticulum-mitochondria crosstalk in patient fibroblasts. Scientific Reports, 2019, 9, 12651.	1.6	28
30	Anandamide Reduces the Toxic Synergism Exerted by Quinolinic Acid and Glutaric Acid in Rat Brain Neuronal Cells. Neuroscience, 2019, 401, 84-95.	1.1	11
31	Acute lysine overload provokes marked striatum injury involving oxidative stress signaling pathways in glutaryl-CoA dehydrogenase deficient mice. Neurochemistry International, 2019, 129, 104467.	1.9	10
32	Pathogenesis of brain damage in glutaric acidemia type I: Lessons from the genetic mice model. International Journal of Developmental Neuroscience, 2019, 78, 215-221.	0.7	17
33	l-Carnitine prevents oxidative stress in striatum of glutaryl-CoA dehydrogenase deficient mice submitted to lysine overload. Biochimica Et Biophysica Acta - Molecular Basis of Disease, 2019, 1865, 2420-2427.	1.8	10
34	Prevention by L-carnitine of DNA damage induced by 3-hydroxy-3-methylglutaric and 3-methylglutaric acids and experimental evidence of lipid and DNA damage in patients with 3-hydroxy-3-methylglutaric aciduria. Archives of Biochemistry and Biophysics, 2019, 668, 16-22.	1.4	5
35	Neurological manifestations of organic acidurias. Nature Reviews Neurology, 2019, 15, 253-271.	4.9	43
36	Bezafibrate In Vivo Administration Prevents 3-Methylglutaric Acid-Induced Impairment of Redox Status, Mitochondrial Biogenesis, and Neural Injury in Brain of Developing Rats. Neurotoxicity Research, 2019, 35, 809-822.	1.3	12

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37	Mitochondrial energetics is impaired in very long-chain acyl-CoA dehydrogenase deficiency and can be rescued by treatment with mitochondria-targeted electron scavengers. Human Molecular Genetics, 2019, 28, 928-941.	1.4	41
38	Evidence that thiol group modification and reactive oxygen species are involved in hydrogen sulfide-induced mitochondrial permeability transition pore opening in rat cerebellum. Mitochondrion, 2019, 47, 141-150.	1.6	7
39	The Role of Oxidative Stress and Bioenergetic Dysfunction in Sulfite Oxidase Deficiency: Insights from Animal Models. Neurotoxicity Research, 2019, 35, 484-494.	1.3	22
40	Bezafibrate Prevents Glycine-Induced Increase of Antioxidant Enzyme Activities in Rat Striatum. Molecular Neurobiology, 2019, 56, 29-38.	1.9	10
41	Metabolite accumulation in <scp>VLCAD</scp> deficiency markedly disrupts mitochondrial bioenergetics and Ca²⁺ homeostasis in the heart. FEBS Journal, 2018, 285, 1437-1455.	2.2	19
42	Induction of Neuroinflammatory Response and Histopathological Alterations Caused by Quinolinic Acid Administration in the Striatum of Glutaryl-CoA Dehydrogenase Deficient Mice. Neurotoxicity Research, 2018, 33, 593-606.	1.3	6
43	Experimental Evidence that In Vivo Intracerebral Administration of L-2-Hydroxyglutaric Acid to Neonatal Rats Provokes Disruption of Redox Status and Histopathological Abnormalities in the Brain. Neurotoxicity Research, 2018, 33, 681-692.	1.3	16
44	Evaluation of mitochondrial bioenergetics, dynamics, endoplasmic reticulum-mitochondria crosstalk, and reactive oxygen species in fibroblasts from patients with complex I deficiency. Scientific Reports, 2018, 8, 1165.	1.6	47
45	Glycine Administration Alters MAPK Signaling Pathways and Causes Neuronal Damage in Rat Brain: Putative Mechanisms Involved in the Neurological Dysfunction in Nonketotic Hyperglycinemia. Molecular Neurobiology, 2018, 55, 741-750.	1.9	10
46	S-Adenosylmethionine Promotes Oxidative Stress and Decreases Na+, K+-ATPase Activity in Cerebral Cortex Supernatants of Adolescent Rats: Implications for the Pathogenesis of S-Adenosylhomocysteine Hydrolase Deficiency. Molecular Neurobiology, 2018, 55, 5868-5878.	1.9	9
47	Toxic Synergism Between Quinolinic Acid and Glutaric Acid in Neuronal Cells Is Mediated by Oxidative Stress: Insights to a New Toxic Model. Molecular Neurobiology, 2018, 55, 5362-5376.	1.9	11
48	Chronic Exposure to Î²-Alanine Generates Oxidative Stress and Alters Energy Metabolism in Cerebral Cortex and Cerebellum of Wistar Rats. Molecular Neurobiology, 2018, 55, 5101-5110.	1.9	19
49	Evidence that Thiosulfate Inhibits Creatine Kinase Activity in Rat Striatum via Thiol Group Oxidation. Neurotoxicity Research, 2018, 34, 693-705.	1.3	18
50	Oxidative damage in glutaric aciduria type I patients and the protective effects of l â€carnitine treatment. Journal of Cellular Biochemistry, 2018, 119, 10021-10032.	1.2	30
51	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. Toxicology in Vitro, 2017, 42, 47-53.	1.1	26
52	Disruption of Energy Transfer and Redox Status by Sulfite in Hippocampus, Striatum, and Cerebellum of Developing Rats. Neurotoxicity Research, 2017, 32, 264-275.	1.3	11
53	Î±-Ketoadipic Acid and Î±-Aminoadipic Acid Cause Disturbance of Glutamatergic Neurotransmission and Induction of Oxidative Stress In Vitro in Brain of Adolescent Rats. Neurotoxicity Research, 2017, 32, 276-290.	1.3	15
54	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. Biochimica Et Biophysica Acta - Molecular Basis of Disease, 2017, 1863, 2135-2148.	1.8	42

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55	Mevalonolactone disrupts mitochondrial functions and induces permeability transition pore opening in rat brain mitochondria: Implications for the pathogenesis of mevalonic aciduria. Neurochemistry International, 2017, 108, 133-145.	1.9	8
56	Impairment of <scp>GABA</scp>ergic system contributes to epileptogenesis in glutaric acidemia type I. Epilepsia, 2017, 58, 1771-1781.	2.6	12
57	Higher Vulnerability of Menadione-Exposed Cortical Astrocytes of Glutaryl-CoA Dehydrogenase Deficient Mice to Oxidative Stress, Mitochondrial Dysfunction, and Cell Death: Implications for the Neurodegeneration in Glutaric Aciduria Type I. Molecular Neurobiology, 2017, 54, 4795-4805.	1.9	7
58	2â€Methylcitric acid impairs glutamate metabolism and induces permeability transition in brain mitochondria. Journal of Neurochemistry, 2016, 137, 62-75.	2.1	27
59	Mitochondrial dysfunction in fatty acid oxidation disorders: insights from human and animal studies. Bioscience Reports, 2016, 36, e00281.	1.1	138
60	3-Hydroxy-3-methylglutaric and 3-methylglutaric acids impair redox status and energy production and transfer in rat heart: relevance for the pathophysiology of cardiac dysfunction in 3-hydroxy-3-methylglutaryl-coenzyme A lyase deficiency. Free Radical Research, 2016, 50, 997-1010.	1.5	19
61	Ornithine and Homocitrulline Impair Mitochondrial Function, Decrease Antioxidant Defenses and Induce Cell Death in Menadione-Stressed Rat Cortical Astrocytes: Potential Mechanisms of Neurological Dysfunction in HHH Syndrome. Neurochemical Research, 2016, 41, 2190-2198.	1.6	14
62	Experimental Evidence that 3-Methylglutaric Acid Disturbs Mitochondrial Function and Induced Oxidative Stress in Rat Brain Synaptosomes: New Converging Mechanisms. Neurochemical Research, 2016, 41, 2619-2626.	1.6	15
63	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. Molecular Neurobiology, 2016, 53, 6459-6475.	1.9	35
64	Induction of a Proinflammatory Response in Cortical Astrocytes by the Major Metabolites Accumulating in HMG-CoA Lyase Deficiency: the Role of ERK Signaling Pathway in Cytokine Release. Molecular Neurobiology, 2016, 53, 3586-3595.	1.9	15
65	Intracerebral Glycine Administration Impairs Energy and Redox Homeostasis and Induces Glial Reactivity in Cerebral Cortex of Newborn Rats. Molecular Neurobiology, 2016, 53, 5864-5875.	1.9	16
66	Deregulation of mitochondrial functions provoked by longâ€chain fatty acid accumulating in longâ€chain 3â€hydroxyacylâ€CoA dehydrogenase and mitochondrial permeability transition deficiencies in rat heart â€“ mitochondrial permeability transition pore opening as a potential contributing pathomechanism of cardiac alterations in these disorders. FEBS Journal, 2015, 282, 4714-4726.	2.2	17
67	Investigation of inflammatory profile in MSUD patients: benefit of L-carnitine supplementation. Metabolic Brain Disease, 2015, 30, 1167-1174.	1.4	29
68	Striatal neuronal death mediated by astrocytes from the Gcdhâˆ’/âˆ’ mouse model of glutaric acidemia type I. Human Molecular Genetics, 2015, 24, 4504-4515.	1.4	25
69	Experimental evidence that bioenergetics disruption is not mainly involved in the brain injury of glutaryl-CoA dehydrogenase deficient mice submitted to lysine overload. Brain Research, 2015, 1620, 116-129.	1.1	13
70	Disturbance of redox homeostasis as a contributing underlying pathomechanism of brain and liver alterations in 3â€hydroxyâ€3â€methylglutarylâ€CoA lyase deficiency. Journal of Inherited Metabolic Disease, 2015, 38, 1021-1028.	1.7	18
71	Disturbance of energy and redox homeostasis and reduction of Na+,K+-ATPase activity provoked by in vivo intracerebral administration of ethylmalonic acid to young rats. Biochimica Et Biophysica Acta - Molecular Basis of Disease, 2015, 1852, 759-767.	1.8	16
72	Increased oxidative stress in patients with 3-hydroxy-3-methylglutaric aciduria. Molecular and Cellular Biochemistry, 2015, 402, 149-155.	1.4	25

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73	NMDA Receptors and Oxidative Stress Induced by the Major Metabolites Accumulating in HMG Lyase Deficiency Mediate Hypophosphorylation of Cytoskeletal Proteins in Brain From Adolescent Rats: Potential Mechanisms Contributing to the Neuropathology of This Disease. Neurotoxicity Research, 2015, 28, 239-252.	1.3	7
74	Reactive nitrogen species mediate oxidative stress and astrogliosis provoked by in vivo administration of phytanic acid in cerebellum of adolescent rats: A potential contributing pathomechanism of cerebellar injury in peroxisomal disorders. Neuroscience, 2015, 304, 122-132.	1.1	22
75	In vivo intracerebral administration of L-2-hydroxyglutaric acid provokes oxidative stress and histopathological alterations in striatum and cerebellum of adolescent rats. Free Radical Biology and Medicine, 2015, 83, 201-213.	1.3	24
76	Ornithine In Vivo Administration Disrupts Redox Homeostasis and Decreases Synaptic Na+, K+-ATPase Activity in Cerebellum of Adolescent Rats: Implications for the Pathogenesis of Hyperornithinemia-Hyperammonemia-Homocitrullinuria (HHH) Syndrome. Cellular and Molecular Neurobiology, 2015, 35, 797-806.	1.7	4
77	Evidence that 3-hydroxy-3-methylglutaric and 3-methylglutaric acids induce DNA damage in rat striatum. Metabolic Brain Disease, 2015, 30, 1055-1062.	1.4	8
78	Toxic synergism between quinolinic acid and organic acids accumulating in glutaric acidemia type I and in disorders of propionate metabolism in rat brain synaptosomes: Relevance for metabolic acidemias. Neuroscience, 2015, 308, 64-74.	1.1	23
79	The effect of WIN 55,212-2 suggests a cannabinoid-sensitive component in the early toxicity induced by organic acids accumulating in glutaric acidemia type I and in related disorders of propionate metabolism in rat brain synaptosomes. Neuroscience, 2015, 310, 578-588.	1.1	14
80	Experimental evidence that overexpression of NR2B glutamate receptor subunit is associated with brain vacuolation in adult glutaryl-CoA dehydrogenase deficient mice: A potential role for glutamatergic-induced excitotoxicity in GA I neuropathology. Journal of the Neurological Sciences, 2015, 359, 133-140.	0.3	14
81	Pristanic Acid Provokes Lipid, Protein, and DNA Oxidative Damage and Reduces the Antioxidant Defenses in Cerebellum of Young Rats. Cerebellum, 2014, 13, 751-759.	1.4	8
82	Homocysteine contribution to DNA damage in cystathionine Î²-synthase-deficient patients. Gene, 2014, 539, 270-274.	1.0	17
83	Ethylmalonic Acid Induces Permeability Transition in Isolated Brain Mitochondria. Neurotoxicity Research, 2014, 26, 168-178.	1.3	11
84	Sulfite disrupts brain mitochondrial energy homeostasis and induces mitochondrial permeability transition pore opening via thiol group modification. Biochimica Et Biophysica Acta - Molecular Basis of Disease, 2014, 1842, 1413-1422.	1.8	31
85	l-carnitine supplementation as a potential antioxidant therapy for inherited neurometabolic disorders. Gene, 2014, 533, 469-476.	1.0	180
86	Disturbance of the glutamatergic system by glutaric acid in striatum and cerebral cortex of glutaryl-CoA dehydrogenase-deficient knockout mice: Possible implications for the neuropathology of glutaric acidemia type I. Journal of the Neurological Sciences, 2014, 346, 260-267.	0.3	11
87	Acute lysine overload provokes protein oxidative damage and reduction of antioxidant defenses in the brain of infant glutaryl-CoA dehydrogenase deficient mice: A role for oxidative stress in GA I neuropathology. Journal of the Neurological Sciences, 2014, 344, 105-113.	0.3	14
88	Disruption of redox homeostasis and histopathological alterations caused by in vivo intrastriatal administration of D-2-hydroxyglutaric acid to young rats. Neuroscience, 2014, 277, 281-293.	1.1	12
89	Evidence that glycine induces lipid peroxidation and decreases glutathione concentrations in rat cerebellum. Molecular and Cellular Biochemistry, 2014, 395, 125-134.	1.4	9
90	Disruption of oxidative phosphorylation and synaptic Na+, K+-ATPase activity by pristanic acid in cerebellum of young rats. Life Sciences, 2014, 94, 67-73.	2.0	0

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91	Mitochondrial bioenergetics deregulation caused by long-chain 3-hydroxy fatty acids accumulating in LCHAD and MTP deficiencies in rat brain: A possible role of mPTP opening as a pathomechanism in these disorders?. Biochimica Et Biophysica Acta - Molecular Basis of Disease, 2014, 1842, 1658-1667.	1.8	22
92	Increased Glutamate Receptor and Transporter Expression in the Cerebral Cortex and Striatum of Gcdh-/- Mice: Possible Implications for the Neuropathology of Glutaric Acidemia Type I. PLoS ONE, 2014, 9, e90477.	1.1	22
93	Glycine Intracerebroventricular Administration Disrupts Mitochondrial Energy Homeostasis in Cerebral Cortex and Striatum of Young Rats. Neurotoxicity Research, 2013, 24, 502-511.	1.3	12
94	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. Journal of Bioenergetics and Biomembranes, 2013, 45, 47-57.	1.0	39
95	Marked inhibition of Na+, K+ - ATPase activity and the respiratory chain by phytanic acid in cerebellum from young rats: possible underlying mechanisms of cerebellar ataxia in Refsum disease. Journal of Bioenergetics and Biomembranes, 2013, 45, 137-144.	1.0	14
96	Neurodevelopmental and cognitive behavior of glutaryl-CoA dehydrogenase deficient knockout mice. Life Sciences, 2013, 92, 137-142.	2.0	10
97	Disturbance of brain energy and redox homeostasis provoked by sulfite and thiosulfate: Potential pathomechanisms involved in the neuropathology of sulfite oxidase deficiency. Gene, 2013, 531, 191-198.	1.0	35
98	Redox homeostasis is compromised in vivo by the metabolites accumulating in 3-hydroxy-3-methylglutaryl-CoA lyase deficiency in rat cerebral cortex and liver. Free Radical Research, 2013, 47, 1066-1075.	1.5	21
99	Disruption of brain redox homeostasis in glutaryl-CoA dehydrogenase deficient mice treated with high dietary lysine supplementation. Molecular Genetics and Metabolism, 2013, 108, 30-39.	0.5	29
100	Neurochemical Evidence that the Metabolites Accumulating in 3-Methylcrotonyl-CoA Carboxylase Deficiency Induce Oxidative Damage in Cerebral Cortex of Young Rats. Cellular and Molecular Neurobiology, 2013, 33, 137-146.	1.7	13
101	In vivo experimental evidence that the major metabolites accumulating in 3-hydroxy-3-methylglutaryl-CoA lyase deficiency induce oxidative stress in striatum of developing rats: A potential pathophysiological mechanism of striatal damage in this disorder. Molecular Genetics and Metabolism, 2013, 109, 144-153.	0.5	23
102	Disturbance of redox homeostasis by ornithine and homocitrulline in rat cerebellum: A possible mechanism of cerebellar dysfunction in HHH syndrome. Life Sciences, 2013, 93, 161-168.	2.0	17
103	Disruption of Mitochondrial Homeostasis by Phytanic Acid in Cerebellum of Young Rats. Cerebellum, 2013, 12, 362-369.	1.4	16
104	Resveratrol Protects C6 Astrocyte Cell Line against Hydrogen Peroxide-Induced Oxidative Stress through Heme Oxygenase 1. PLoS ONE, 2013, 8, e64372.	1.1	114
105	Chronic postnatal ornithine administration to rats provokes learning deficit in the open field task. Metabolic Brain Disease, 2012, 27, 479-486.	1.4	4
106	Impairment of brain redox homeostasis caused by the major metabolites accumulating in hyperornithinemiaâ€“hyperammonemiaâ€“homocitrullinuria syndrome in vivo. Metabolic Brain Disease, 2012, 27, 521-530.	1.4	11
107	Marked reduction of Na+, K+-ATPase and creatine kinase activities induced by acute lysine administration in glutaryl-CoA dehydrogenase deficient mice. Molecular Genetics and Metabolism, 2012, 107, 81-86.	0.5	24
108	Reduction of Na+, K+-ATPase activity and expression in cerebral cortex of glutaryl-CoA dehydrogenase deficient mice: A possible mechanism for brain injury in glutaric aciduria type I. Molecular Genetics and Metabolism, 2012, 107, 375-382.	0.5	24

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109	Ethylmalonic acid impairs brain mitochondrial succinate and malate transport. Molecular Genetics and Metabolism, 2012, 105, 84-90.	0.5	15
110	Induction of oxidative stress in brain of glutaryl-CoA dehydrogenase deficient mice by acute lysine administration. Molecular Genetics and Metabolism, 2012, 106, 31-38.	0.5	29
111	Disruption of redox homeostasis in cerebral cortex of developing rats by acylcarnitines accumulating in mediumâ€chain acylâ€CoA dehydrogenase deficiency. International Journal of Developmental Neuroscience, 2012, 30, 383-390.	0.7	15
112	Phytanic acid disturbs mitochondrial homeostasis in heart of young rats: a possible pathomechanism of cardiomyopathy in Refsum disease. Molecular and Cellular Biochemistry, 2012, 366, 335-343.	1.4	13
113	Experimental evidence that pristanic acid disrupts mitochondrial homeostasis in brain of young rats. Journal of Neuroscience Research, 2012, 90, 597-605.	1.3	8
114	Oxidative Stress Parameters in Urine from Patients with Disorders of Propionate Metabolism: a Beneficial Effect of l-Carnitine Supplementation. Cellular and Molecular Neurobiology, 2012, 32, 77-82.	1.7	30
115	3-Methylcrotonylglycine Disrupts Mitochondrial Energy Homeostasis and Inhibits Synaptic Na+,K+-ATPase Activity in Brain of Young Rats. Cellular and Molecular Neurobiology, 2012, 32, 297-307.	1.7	15
116	Neurochemical evidence that 3â€methylglutaric acid inhibits synaptic Na⁺,K⁺âATPase activity probably through oxidative damage in brain cortex of young rats. International Journal of Developmental Neuroscience, 2011, 29, 1-7.	0.7	27
117	Glycine intrastriatal administration induces lipid and protein oxidative damage and alters the enzymatic antioxidant defenses in rat brain. Life Sciences, 2011, 89, 276-281.	2.0	12
118	Dual mechanism of brain damage induced in vivo by the major metabolites accumulating in hyperornithinemiaâ€“hyperammonemiaâ€“homocitrullinuria syndrome. Brain Research, 2011, 1369, 235-244.	1.1	15
119	Pristanic acid promotes oxidative stress in brain cortex of young rats: A possible pathophysiological mechanism for brain damage in peroxisomal disorders. Brain Research, 2011, 1382, 259-265.	1.1	16
120	Disruption of mitochondrial homeostasis in organic acidurias: insights from human and animal studies. Journal of Bioenergetics and Biomembranes, 2011, 43, 31-38.	1.0	71
121	Mitochondrial energy metabolism in neurodegeneration associated with methylmalonic acidemia. Journal of Bioenergetics and Biomembranes, 2011, 43, 39-46.	1.0	62
122	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
123	Oxidative Stress in Phenylketonuria: What is the Evidence?. Cellular and Molecular Neurobiology, 2011, 31, 653-662.	1.7	67
124	Neurochemical Evidence that Lysine Inhibits Synaptic Na+,K+-ATPase Activity and Provokes Oxidative Damage in Striatum of Young Rats In vivo. Neurochemical Research, 2011, 36, 205-214.	1.6	10
125	Neurochemical Evidence that Pristanic Acid Impairs Energy Production and Inhibits Synaptic Na+, K+-ATPase Activity in Brain of Young Rats. Neurochemical Research, 2011, 36, 1101-1107.	1.6	10
126	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

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127	Induction of S100B secretion in C6 astroglial cells by the major metabolites accumulating in glutaric acidemia type I. Metabolic Brain Disease, 2010, 25, 191-198.	1.4	13
128	Evidence that 2-methylacetoacetate induces oxidative stress in rat brain. Metabolic Brain Disease, 2010, 25, 261-267.	1.4	7
129	Promotion of Lipid and Protein Oxidative Damage in Rat Brain by Ethylmalonic Acid. Neurochemical Research, 2010, 35, 298-305.	1.6	15
130	Î±-Ketoisocaproic acid and leucine provoke mitochondrial bioenergetic dysfunction in rat brain. Brain Research, 2010, 1324, 75-84.	1.1	75
131	In vitro evidence that phytanic acid compromises Na+,K+-ATPase activity and the electron flow through the respiratory chain in brain cortex from young rats. Brain Research, 2010, 1352, 231-238.	1.1	24
132	An overview of L-2-hydroxyglutarate dehydrogenase gene (L2HGDH) variants: a genotype-phenotype study. Human Mutation, 2010, 31, 380-390.	1.1	108
133	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
134	Neurochemical evidence that glycine induces bioenergetical dysfunction. Neurochemistry International, 2010, 56, 948-954.	1.9	21
135	Reduction of lipid and protein damage in patients with disorders of propionate metabolism under treatment: a possible protective role of l â€carnitine supplementation. International Journal of Developmental Neuroscience, 2010, 28, 127-132.	0.7	45
136	<scp>d</scp>â€Serine administration provokes lipid oxidation and decreases the antioxidant defenses in rat striatum. International Journal of Developmental Neuroscience, 2010, 28, 297-301.	0.7	9
137	Disturbance of mitochondrial energy homeostasis caused by the metabolites accumulating in LCHAD and MTP deficiencies in rat brain. Life Sciences, 2010, 86, 825-831.	2.0	30
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