

Carlo Viscomi

List of Publications by Year in descending order

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Version: 2024-02-01

83
papers

6,635
citations

71102

41
h-index

64796

79
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93
all docs

93
docs citations

93
times ranked

9470
citing authors

#	ARTICLE	IF	CITATIONS
1	MPV17 encodes an inner mitochondrial membrane protein and is mutated in infantile hepatic mitochondrial DNA depletion. <i>Nature Genetics</i> , 2006, 38, 570-575.	21.4	380
2	Loss of ETHE1, a mitochondrial dioxygenase, causes fatal sulfide toxicity in ethylmalonic encephalopathy. <i>Nature Medicine</i> , 2009, 15, 200-205.	30.7	358
3	The Opa1-Dependent Mitochondrial Cristae Remodeling Pathway Controls Atrophic, Apoptotic, and Ischemic Tissue Damage. <i>Cell Metabolism</i> , 2015, 21, 834-844.	16.2	350
4	Niche stiffness underlies the ageing of central nervous system progenitor cells. <i>Nature</i> , 2019, 573, 130-134.	27.8	311
5	NAD ⁺ -Dependent Activation of Sirt1 Corrects the Phenotype in a Mouse Model of Mitochondrial Disease. <i>Cell Metabolism</i> , 2014, 19, 1042-1049.	16.2	293
6	Transcription Factor EB Controls Metabolic Flexibility during Exercise. <i>Cell Metabolism</i> , 2017, 25, 182-196.	16.2	250
7	In Vivo Correction of COX Deficiency by Activation of the AMPK/PGC-1 β Axis. <i>Cell Metabolism</i> , 2011, 14, 80-90.	16.2	245
8	Deep bradycardia and heart block caused by inducible cardiac-specific knockout of the pacemaker channel gene <i>Hcn4</i> . <i>Proceedings of the National Academy of Sciences of the United States of America</i> , 2011, 108, 1705-1710.	7.1	240
9	Genome editing in mitochondria corrects a pathogenic mtDNA mutation in vivo. <i>Nature Medicine</i> , 2018, 24, 1691-1695.	30.7	215
10	Pharmacological Inhibition of Poly(ADP-Ribose) Polymerases Improves Fitness and Mitochondrial Function in Skeletal Muscle. <i>Cell Metabolism</i> , 2014, 19, 1034-1041.	16.2	211
11	Control of mitochondrial superoxide production by reverse electron transport at complex I. <i>Journal of Biological Chemistry</i> , 2018, 293, 9869-9879.	3.4	204
12	Opa1 Overexpression Ameliorates the Phenotype of Two Mitochondrial Disease Mouse Models. <i>Cell Metabolism</i> , 2015, 21, 845-854.	16.2	202
13	Cryo-EM structures of complex I from mouse heart mitochondria in two biochemically defined states. <i>Nature Structural and Molecular Biology</i> , 2018, 25, 548-556.	8.2	202
14	Heteromeric HCN1/HCN4 Channels: A Comparison with Native Pacemaker Channels from the Rabbit Sinoatrial Node. <i>Journal of Physiology</i> , 2003, 549, 347-359.	2.9	185
15	FGF21 is a biomarker for mitochondrial translation and mtDNA maintenance disorders. <i>Neurology</i> , 2016, 87, 2290-2299.	1.1	167
16	MtDNA maintenance defects: syndromes and genes. <i>Journal of Inherited Metabolic Disease</i> , 2017, 40, 587-599.	3.6	145
17	Integrated Allosteric Model of Voltage Gating of Hcn Channels. <i>Journal of General Physiology</i> , 2001, 117, 519-532.	1.9	144
18	Combined treatment with oral metronidazole and N-acetylcysteine is effective in ethylmalonic encephalopathy. <i>Nature Medicine</i> , 2010, 16, 869-871.	30.7	136

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19	Emerging concepts in the therapy of mitochondrial disease. <i>Biochimica Et Biophysica Acta - Bioenergetics</i> , 2015, 1847, 544-557.	1.0	96
20	Neural stem cells traffic functional mitochondria via extracellular vesicles. <i>PLoS Biology</i> , 2021, 19, e3001166.	5.6	95
21	Interaction of the Pacemaker Channel HCN1 with Filamin A. <i>Journal of Biological Chemistry</i> , 2004, 279, 43847-43853.	3.4	91
22	Complex IV-deficient <i>Surf1</i> mice initiate mitochondrial stress responses. <i>Biochemical Journal</i> , 2014, 462, 359-371.	3.7	89
23	Early-onset liver mtDNA depletion and late-onset proteinuric nephropathy in <i>Mpv17</i> knockout mice. <i>Human Molecular Genetics</i> , 2009, 18, 12-26.	2.9	87
24	Rapamycin rescues mitochondrial myopathy via coordinated activation of autophagy and lysosomal biogenesis. <i>EMBO Molecular Medicine</i> , 2018, 10, .	6.9	86
25	Chronic Exposure to Sulfide Causes Accelerated Degradation of Cytochrome c Oxidase in Ethylmalonic Encephalopathy. <i>Antioxidants and Redox Signaling</i> , 2011, 15, 353-362.	5.4	80
26	Loss of function of the mitochondrial peptidase PITRM1 induces proteotoxic stress and Alzheimer's disease-like pathology in human cerebral organoids. <i>Molecular Psychiatry</i> , 2021, 26, 5733-5750.	7.9	79
27	Effective AAV-mediated gene therapy in a mouse model of ethylmalonic encephalopathy. <i>EMBO Molecular Medicine</i> , 2012, 4, 1008-1014.	6.9	72
28	Structural basis for a complex I mutation that blocks pathological ROS production. <i>Nature Communications</i> , 2021, 12, 707.	12.8	71
29	Paroxysmal non-kinesigenic dyskinesia is caused by mutations of the MR-1 mitochondrial targeting sequence. <i>Human Molecular Genetics</i> , 2009, 18, 1058-1064.	2.9	70
30	Localized cerebral energy failure in DNA polymerase gamma-associated encephalopathy syndromes. <i>Brain</i> , 2010, 133, 1428-1437.	7.6	70
31	Perturbed Redox Signaling Exacerbates a Mitochondrial Myopathy. <i>Cell Metabolism</i> , 2018, 28, 764-775.e5.	16.2	70
32	TTC19 Plays a Husbandry Role on UQCRC1 Turnover in the Biogenesis of Mitochondrial Respiratory Complex III. <i>Molecular Cell</i> , 2017, 67, 96-105.e4.	9.7	64
33	Defective PITRM1 mitochondrial peptidase is associated with A β amyloidotic neurodegeneration. <i>EMBO Molecular Medicine</i> , 2016, 8, 176-190.	6.9	60
34	C Terminus-mediated Control of Voltage and cAMP Gating of Hyperpolarization-activated Cyclic Nucleotide-gated Channels. <i>Journal of Biological Chemistry</i> , 2001, 276, 29930-29934.	3.4	58
35	Gene Therapy Using a Liver-targeted AAV Vector Restores Nucleoside and Nucleotide Homeostasis in a Murine Model of MNGIE. <i>Molecular Therapy</i> , 2014, 22, 901-907.	8.2	55
36	Kinetic and ionic properties of the human HCN2 pacemaker channel. <i>Pflugers Archiv European Journal of Physiology</i> , 2000, 439, 618-626.	2.8	52

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37	Evolution Meets Disease: Penetrance and Functional Epistasis of Mitochondrial tRNA Mutations. <i>PLoS Genetics</i> , 2011, 7, e1001379.	3.5	51
38	AAV9-based gene therapy partially ameliorates the clinical phenotype of a mouse model of Leigh syndrome. <i>Gene Therapy</i> , 2017, 24, 661-667.	4.5	50
39	AAV-mediated Liver-specific MPV17 Expression Restores mtDNA Levels and Prevents Diet-induced Liver Failure. <i>Molecular Therapy</i> , 2014, 22, 10-17.	8.2	47
40	Strategies for fighting mitochondrial diseases. <i>Journal of Internal Medicine</i> , 2020, 287, 665-684.	6.0	47
41	Towards a therapy for mitochondrial disease: an update. <i>Biochemical Society Transactions</i> , 2018, 46, 1247-1261.	3.4	46
42	Bioenergetic consequences from xenotopic expression of a tunicate AOX in mouse mitochondria: Switch from RET and ROS to FET. <i>Biochimica Et Biophysica Acta - Bioenergetics</i> , 2020, 1861, 148137.	1.0	46
43	Metabolic effects of bezafibrate in mitochondrial disease. <i>EMBO Molecular Medicine</i> , 2020, 12, e11589.	6.9	45
44	The short N-terminus is required for functional expression of the virus-encoded miniature K ⁺ channel Kcv. <i>FEBS Letters</i> , 2002, 530, 65-69.	2.8	39
45	Long-Term Sustained Effect of Liver-Targeted Adeno-Associated Virus Gene Therapy for Mitochondrial Neurogastrointestinal Encephalomyopathy. <i>Human Gene Therapy</i> , 2018, 29, 708-718.	2.7	39
46	Effects of dronedarone on Acetylcholine-activated current in rabbit SAN cells. <i>British Journal of Pharmacology</i> , 2000, 130, 1315-1320.	5.4	37
47	Kinetic and ionic properties of the human HCN2 pacemaker channel. <i>Pflügers Archiv European Journal of Physiology</i> , 2000, 439, 618-626.	2.8	37
48	Morphologic evidence of diffuse vascular damage in human and in the experimental model of ethylmalonic encephalopathy. <i>Journal of Inherited Metabolic Disease</i> , 2012, 35, 451-458.	3.6	35
49	Decreased <i>in vitro</i> Mitochondrial Function is Associated with Enhanced Brain Metabolism, Blood Flow, and Memory in Surf1-Deficient Mice. <i>Journal of Cerebral Blood Flow and Metabolism</i> , 2013, 33, 1605-1611.	4.3	35
50	A Single Intravenous Injection of AAV-PHP.B-hNDUFS4 Ameliorates the Phenotype of Ndufs4 Mice. <i>Molecular Therapy - Methods and Clinical Development</i> , 2020, 17, 1071-1078.	4.1	32
51	Proteome adaptations in Ethe1-deficient mice indicate a role in lipid catabolism and cytoskeleton organization via post-translational protein modifications. <i>Bioscience Reports</i> , 2013, 33, .	2.4	31
52	Improved insulin sensitivity associated with reduced mitochondrial complex IV assembly and activity. <i>FASEB Journal</i> , 2013, 27, 1371-1380.	0.5	29
53	Defective endoplasmic reticulum-mitochondria contacts and bioenergetics in SEP1N-related myopathy. <i>Cell Death and Differentiation</i> , 2021, 28, 123-138.	11.2	29
54	Mitochondrial Neurodegeneration. <i>Cells</i> , 2022, 11, 637.	4.1	29

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55	Physical and Functional Cross Talk Between Endo-Sarcoplasmic Reticulum and Mitochondria in Skeletal Muscle. <i>Antioxidants and Redox Signaling</i> , 2020, 32, 873-883.	5.4	27
56	Toward a therapy for mitochondrial disease. <i>Biochemical Society Transactions</i> , 2016, 44, 1483-1490.	3.4	26
57	SURF1 knockout cloned pigs: Early onset of a severe lethal phenotype. <i>Biochimica Et Biophysica Acta - Molecular Basis of Disease</i> , 2018, 1864, 2131-2142.	3.8	24
58	Down-regulation of the mitochondrial aspartate-glutamate carrier isoform 1 AGC1 inhibits proliferation and N-acetylaspartate synthesis in Neuro2A cells. <i>Biochimica Et Biophysica Acta - Molecular Basis of Disease</i> , 2017, 1863, 1422-1435.	3.8	22
59	Mitochondrial DNA heteroplasmy is modulated during oocyte development propagating mutation transmission. <i>Science Advances</i> , 2021, 7, eabi5657.	10.3	22
60	Tissue- and species-specific differences in cytochrome c oxidase assembly induced by SURF1 defects. <i>Biochimica Et Biophysica Acta - Molecular Basis of Disease</i> , 2016, 1862, 705-715.	3.8	21
61	APOPT 1/ COA 8 assists COX assembly and is oppositely regulated by UPS and ROS. <i>EMBO Molecular Medicine</i> , 2019, 11, .	6.9	19
62	Role of PITRM1 in Mitochondrial Dysfunction and Neurodegeneration. <i>Biomedicines</i> , 2021, 9, 833.	3.2	17
63	A novel heteroplasmic tRNASer(UCN) mtDNA point mutation associated with progressive external ophthalmoplegia and hearing loss. <i>Neuromuscular Disorders</i> , 2007, 17, 681-683.	0.6	16
64	Exploiting pyocyanin to treat mitochondrial disease due to respiratory complex III dysfunction. <i>Nature Communications</i> , 2021, 12, 2103.	12.8	16
65	DNA polymerase gamma mutations that impair holoenzyme stability cause catalytic subunit depletion. <i>Nucleic Acids Research</i> , 2021, 49, 5230-5248.	14.5	15
66	Respiratory chain signalling is essential for adaptive remodelling following cardiac ischaemia. <i>Journal of Cellular and Molecular Medicine</i> , 2020, 24, 3534-3548.	3.6	15
67	Lifelong reduction in complex IV induces tissue-specific metabolic effects but does not reduce lifespan or healthspan in mice. <i>Aging Cell</i> , 2018, 17, e12769.	6.7	14
68	Quantitative proteomics suggests metabolic reprogramming during ETHE1 deficiency. <i>Proteomics</i> , 2016, 16, 1166-1176.	2.2	12
69	Hypoxic and hypercapnic challenges unveil respiratory vulnerability of Surf1 knockout mice, an animal model of Leigh syndrome. <i>Mitochondrion</i> , 2011, 11, 413-420.	3.4	11
70	Opa1 Overexpression Protects from Early-Onset Mpv17-Related Mouse Kidney Disease. <i>Molecular Therapy</i> , 2020, 28, 1918-1930.	8.2	9
71	CG7630 is the <i>Drosophila melanogaster</i> homolog of the cytochrome c oxidase subunit COX7B. <i>EMBO Reports</i> , 0, .	4.5	7
72	Lactic Acidosis in a Newborn With Adrenal Calcifications. <i>Pediatric Research</i> , 2009, 66, 317-322.	2.3	5

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73	The homeostatic dynamics of feeding behaviour identify novel mechanisms of anorectic agents. PLoS Biology, 2019, 17, e3000482.	5.6	5
74	Redox Signaling and Stress in Inherited Myopathies. Antioxidants and Redox Signaling, 2022, 37, 301-323.	5.4	5
75	Mitochondrial Cytochrome c Oxidase Defects Alter Cellular Homeostasis of Transition Metals. Frontiers in Cell and Developmental Biology, 2022, 10, .	3.7	5
76	Effects of ketosis in mitochondrial myopathy: potential benefits of a mitotoxic diet. EMBO Molecular Medicine, 2016, 8, 1231-1233.	6.9	4
77	Breathe: Your Mitochondria Will Do the Rest! If They Are Healthy!. Cell Metabolism, 2019, 30, 628-629.	16.2	4
78	Response to Drs. Djouadi and Bastin. Cell Metabolism, 2011, 14, 717.	16.2	1
79	Data on cytochrome c oxidase assembly in mice and human fibroblasts or tissues induced by SURF1 defect. Data in Brief, 2016, 7, 1004-1009.	1.0	1
80	Mitochondrial Genes and Neurodegenerative Disease. , 2016, , 81-106.		1
81	Molecular Research on Mitochondrial Dysfunction. International Journal of Molecular Sciences, 2022, 23, 6845.	4.1	1
82	Reply to Dr Michaudet. European Journal of Neurology, 2015, 22, e78-e78.	3.3	0
83	Experimental Therapies. , 2019, , 357-370.		0