Pierre Rustin

List of Publications by Year
in descending order

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216 papers	24,382 citations	6592 79 h-index	7496 151 g-index
223 all docs	223 docs citations	223 times ranked	21589 citing authors

#	Article	IF	Citations
1	Mitochondrial transcription factor A is necessary for mtDNA maintance and embryogenesis in mice. Nature Genetics, 1998, 18, 231-236.	9.4	1,377
2	Aconitase and mitochondrial ironâ€“sulphur protein deficiency in Friedreich ataxia. Nature Genetics, 1997, 17, 215-217.	9.4	1,027
3	Mitochondrial transcription factor A regulates mtDNA copy number in mammals. Human Molecular Genetics, 2004, 13, 935-944.	1.4	730
4	Mouse models for Friedreich ataxia exhibit cardiomyopathy, sensory nerve defect and Fe-S enzyme deficiency followed by intramitochondrial iron deposits. Nature Genetics, 2001, 27, 181-186.	9.4	700
5	Mutation of a nuclear succinate dehydrogenase gene results in mitochondrial respiratory chain deficiency. Nature Genetics, 1995, 11, 144-149.	9.4	684
6	Persistent mitochondrial dysfunction and perinatal exposure to antiretroviral nucleoside analogues. Lancet, The, 1999, 354, 1084-1089.	6.3	627
7	SDH Mutations Establish a Hypermethylator Phenotype in Paraganglioma. Cancer Cell, 2013, 23, 739-752.	7.7	606
8	SDHA is a tumor suppressor gene causing paraganglioma. Human Molecular Genetics, 2010, 19, 3011-3020.	1.4	604
9	AIF deficiency compromises oxidative phosphorylation. EMBO Journal, 2004, 23, 4679-4689.	3.5	576
10	Defects in succinate dehydrogenase in gastrointestinal stromal tumors lacking KIT and PDGFRA mutations. Proceedings of the National Academy of Sciences of the United States of America, 2011, 108, 314-318.	3.3	574
11	Clinical and molecular genetics of patients with the Carneyâ€“Stratakis syndrome and germline mutations of the genes coding for the succinate dehydrogenase subunits SDHB, SDHC, and SDHD. European Journal of Human Genetics, 2008, 16, 79-88.	1.4	446
12	Spermidine and resveratrol induce autophagy by distinct pathways converging on the acetylproteome. Journal of Cell Biology, 2011, 192, 615-629.	2.3	439
13	Mutations in the SDHB gene are associated with extra-adrenal and/or malignant phaeochromocytomas. Cancer Research, 2003, 63, 5615-21.	0.4	409
14	Imbalanced OPA1 processing and mitochondrial fragmentation cause heart failure in mice. Science, 2015, 350, aad0116.	6.0	403
15	Dilated cardiomyopathy and atrioventricular conduction blocks induced by heart-specific inactivation of mitochondrial DNA gene expression. Nature Genetics, 1999, 21, 133-137.	9.4	393
16	Targeted Deletion of AIF Decreases Mitochondrial Oxidative Phosphorylation and Protects from Obesity and Diabetes. Cell, 2007, 131, 476-491.	13.5	381
17	Effect of idebenone on cardiomyopathy in Friedreich's ataxia: a preliminary study. Lancet, The, 1999, 354, 477-479.	6.3	352
18	Quinone-responsive multiple respiratory-chain dysfunction due to widespread coenzyme Q10 deficiency. Lancet, The, 2000, 356, 391-395.	6.3	349

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19	The R22X Mutation of the SDHD Gene in Hereditary Paraganglioma Abolishes the Enzymatic Activity of Complex II in the Mitochondrial Respiratory Chain and Activates the Hypoxia Pathway. American Journal of Human Genetics, 2001, 69, 1186-1197.	2.6	339
20	Mutations of theSCO1Gene in Mitochondrial CytochromecOxidase Deficiency with Neonatalâ€Onset Hepatic Failure and Encephalopathy. American Journal of Human Genetics, 2000, 67, 1104-1109.	2.6	322
21	Germline mutations in FH confer predisposition to malignant pheochromocytomas and paragangliomas. Human Molecular Genetics, 2014, 23, 2440-2446.	1.4	316
22	A mutant mitochondrial respiratory chain assembly protein causes complex III deficiency in patients with tubulopathy, encephalopathy and liver failure. Nature Genetics, 2001, 29, 57-60.	9.4	297
23	Mitochondria are physiologically maintained at close to 50 Â°C. PLoS Biology, 2018, 16, e2003992.	2.6	295
24	A mutation in the human heme A:farnesyltransferase gene (COX10) causes cytochrome c oxidase deficiency. Human Molecular Genetics, 2000, 9, 1245-1249.	1.4	261
25	Control of Mitochondrial Membrane Permeabilization by Adenine Nucleotide Translocator Interacting with HIV-1 Viral Protein R and Bcl-2. Journal of Experimental Medicine, 2001, 193, 509-520.	4.2	261
26	Persistent mitochondrial dysfunction in HIV-1-exposed but uninfected infants. Aids, 2003, 17, 1769-1785.	1.0	251
27	Large-Scale Deletion and Point Mutations of the Nuclear NDUFV1 and NDUFS1 Genes in Mitochondrial Complex I Deficiency. American Journal of Human Genetics, 2001, 68, 1344-1352.	2.6	243
28	Clinical spectrum and diagnosis of mitochondrial disorders. American Journal of Medical Genetics Part A, 2001, 106, 4-17.	2.4	215
29	Functional Consequences of aSDHBGene Mutation in an Apparently Sporadic Pheochromocytoma. Journal of Clinical Endocrinology and Metabolism, 2002, 87, 4771-4774.	1.8	210
30	Succinate dehydrogenase and human diseases: new insights into a well-known enzyme. European Journal of Human Genetics, 2002, 10, 289-291.	1.4	208
31	Muscle-Specific Loss of Apoptosis-Inducing Factor Leads to Mitochondrial Dysfunction, Skeletal Muscle Atrophy, and Dilated Cardiomyopathy. Molecular and Cellular Biology, 2005, 25, 10261-10272.	1.1	208
32	The Warburg Effect Is Genetically Determined in Inherited Pheochromocytomas. PLoS ONE, 2009, 4, e7094.	1.1	203
33	The Human Cytochrome c Oxidase Assembly Factors SCO1 and SCO2 Have Regulatory Roles in the Maintenance of Cellular Copper Homeostasis. Cell Metabolism, 2007, 5, 9-20.	7.2	197
34	Spectrum of mitochondrial DNA rearrangements in the Pearson marrow-pancreas syndrome. Human Molecular Genetics, 1995, 4, 1327-1330.	1.4	193
35	Friedreich Ataxia Mouse Models with Progressive Cerebellar and Sensory Ataxia Reveal Autophagic Neurodegeneration in Dorsal Root Ganglia. Journal of Neuroscience, 2004, 24, 1987-1995.	1.7	189
36	Impaired Nuclear Nrf2 Translocation Undermines the Oxidative Stress Response in Friedreich Ataxia. PLoS ONE, 2009, 4, e4253.	1.1	181

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37	Disabled early recruitment of antioxidant defenses in Friedreich's ataxia. Human Molecular Genetics, 2001, 10, 2061-2067.	1.4	176
38	Cytochrome oxidase in health and disease. Gene, 2002, 286, 53-63.	1.0	175
39	Interaction between AIF and CHCHD4 Regulates Respiratory Chain Biogenesis. Molecular Cell, 2015, 58, 1001-1014.	4.5	164
40	S6 Kinase Deletion Suppresses Muscle Growth Adaptations to Nutrient Availability by Activating AMP Kinase. Cell Metabolism, 2007, 5, 476-487.	7.2	163
41	Expression of the Ciona intestinalis Alternative Oxidase (AOX) in Drosophila Complements Defects in Mitochondrial Oxidative Phosphorylation. Cell Metabolism, 2009, 9, 449-460.	7.2	156
42	Stabilization of Hypoxia-inducible Factor-1Î± Protein in Hypoxia Occurs Independently of Mitochondrial Reactive Oxygen Species Production. Journal of Biological Chemistry, 2010, 285, 31277-31284.	1.6	154
43	Mutant NDUFV2 subunit of mitochondrial complex I causes early onset hypertrophic cardiomyopathy and encephalopathy. Human Mutation, 2003, 21, 582-586.	1.1	152
44	Assessment of the mitochondrial respiratory chain. Lancet, The, 1991, 338, 60.	6.3	151
45	Late-Onset Corticohippocampal Neurodepletion Attributable to Catastrophic Failure of Oxidative Phosphorylation in MILON Mice. Journal of Neuroscience, 2001, 21, 8082-8090.	1.7	151
46	Coenzyme Q10 and idebenone in the therapy of respiratory chain diseases: rationale and comparative benefits. Molecular Genetics and Metabolism, 2002, 77, 21-30.	0.5	150
47	Impaired Mitochondrial Glutamate Transport in Autosomal Recessive Neonatal Myoclonic Epilepsy. American Journal of Human Genetics, 2005, 76, 334-339.	2.6	149
48	Mitochondrial succinate is instrumental for HIF1Î± nuclear translocation in SDHA-mutant fibroblasts under normoxic conditions. Human Molecular Genetics, 2005, 14, 3263-3269.	1.4	146
49	Clinical presentations and laboratory investigations in respiratory chain deficiency. European Journal of Pediatrics, 1996, 155, 262-274.	1.3	136
50	Succinate Dehydrogenase (SDH) D Subunit (SDHD) Inactivation in a Growth-Hormone-Producing Pituitary Tumor: A New Association for SDH?. Journal of Clinical Endocrinology and Metabolism, 2012, 97, E357-E366.	1.8	134
51	Expression of the yeast NADH dehydrogenase Ndi1 in Drosophila confers increased lifespan independently of dietary restriction. Proceedings of the National Academy of Sciences of the United States of America, 2010, 107, 9105-9110.	3.3	132
52	A case of mitochondrial encephalomyopathy associated with a muscle coenzyme Q10 deficiency. Journal of the Neurological Sciences, 1998, 156, 41-46.	0.3	129
53	Antenatal manifestations of mitochondrial respiratory chain deficiency. Journal of Pediatrics, 2003, 143, 208-212.	0.9	129
54	A deletion in the human QP-C gene causes a complexÂIII deficiency resulting in hypoglycaemia and lactic acidosis. Human Genetics, 2003, 113, 118-122.	1.8	128

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55	Idebenone delays the onset of cardiac functional alteration without correction of Fe-S enzymes deficit in a mouse model for Friedreich ataxia. Human Molecular Genetics, 2004, 13, 1017-1024.	1.4	128
56	Mitochondrial cytochrome c oxidase deficiency. Clinical Science, 2016, 130, 393-407.	1.8	121
57	Mitochondria, from cell death to proliferation. Nature Genetics, 2002, 30, 352-353.	9.4	118
58	Cytochrome c Oxidase Subassemblies in Fibroblast Cultures from Patients Carrying Mutations in COX10, SCO1, or SURF1. Journal of Biological Chemistry, 2004, 279, 7462-7469.	1.6	118
59	Novel FH mutations in families with hereditary leiomyomatosis and renal cell cancer (HLRCC) and patients with isolated type 2 papillary renal cell carcinoma. Journal of Medical Genetics, 2011, 48, 226-234.	1.5	116
60	Allotopic expression of a mitochondrial alternative oxidase confers cyanide resistance to human cell respiration. EMBO Reports, 2006, 7, 341-345.	2.0	110
61	The Spectrum of Systemic Involvement in Adults Presenting with Renal Lesion and Mitochondrial tRNA(Leu) Gene Mutation. Journal of the American Society of Nephrology: JASN, 2003, 14, 2099-2108.	3.0	109
62	PGC-1Î± is Dispensable for Exercise-Induced Mitochondrial Biogenesis in Skeletal Muscle. PLoS ONE, 2012, 7, e41817.	1.1	108
63	Mitochondrial DNA rearrangements with onset as chronic diarrhea with villous atrophy. Journal of Pediatrics, 1994, 124, 63-70.	0.9	103
64	Absence of Relationship between the Level of Electron Transport Chain Activities and Aging in Human Skeletal Muscle. Biochemical and Biophysical Research Communications, 1996, 229, 536-539.	1.0	97
65	Coamplification of Nuclear Pseudogenes and Assessment of Heteroplasmy of Mitochondrial DNA Mutations. Biochemical and Biophysical Research Communications, 1998, 247, 57-59.	1.0	96
66	Alternative Oxidase Expression in the Mouse Enables Bypassing Cytochrome c Oxidase Blockade and Limits Mitochondrial ROS Overproduction. PLoS Genetics, 2013, 9, e1003182.	1.5	96
67	A mitochondrial cytochrome b mutation but no mutations of nuclearly encoded subunits in ubiquinol cytochrome c reductase (complex III) deficiency. Human Genetics, 1999, 104, 460-466.	1.8	95
68	Tricarboxylic acid cycle dysfunction as a cause of human diseases and tumor formation. American Journal of Physiology - Cell Physiology, 2006, 291, C1114-C1120.	2.1	95
69	Loss of Aif function causes cell death in the mouse embryo, but the temporal progression of patterning is normal. Proceedings of the National Academy of Sciences of the United States of America, 2006, 103, 9918-9923.	3.3	94
70	Disconnecting Mitochondrial Content from Respiratory Chain Capacity in PGC-1-Deficient Skeletal Muscle. Cell Reports, 2013, 3, 1449-1456.	2.9	93
71	Neonatal and delayed-onset liver involvement in disorders of oxidative phosphorylation. Journal of Pediatrics, 1997, 130, 817-822.	0.9	91
72	Cytopathic effects of the cytomegalovirus-encoded apoptosis inhibitory protein vMIA. Journal of Cell Biology, 2006, 174, 985-996.	2.3	90

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73	Expression of the alternative oxidase complements cytochrome c oxidase deficiency in human cells. EMBO Molecular Medicine, 2009, 1, 30-36.	3.3	89
74	Alpha-ketoglutarate dehydrogenase deficiency presenting as congenital lactic acidosis. Journal of Pediatrics, 1992, 121, 255-258.	0.9	88
75	SLC25A32 Mutations and Riboflavin-Responsive Exercise Intolerance. New England Journal of Medicine, 2016, 374, 795-797.	13.9	87
76	Endomyocardial biopsies for early detection of mitochondrial disorders in hypertrophic cardiomyopathies. Journal of Pediatrics, 1994, 124, 224-228.	0.9	86
77	Malate Oxidation in Plant Mitochondria via Malic Enzyme and the Cyanide-insensitive Electron Transport Pathway. Plant Physiology, 1980, 66, 457-462.	2.3	85
78	Three spectrophotometric assays for the measurement of the five respiratory chain complexes in minuscule biological samples. Clinica Chimica Acta, 2006, 374, 81-86.	0.5	83
79	A critical approach to the therapy of mitochondrial respiratory chain and oxidative phosphorylation diseases. Biochimica Et Biophysica Acta - Molecular Basis of Disease, 2009, 1792, 1159-1167.	1.8	82
80	Heart Hypertrophy and Function Are Improved by Idebenone in Friedreich's Ataxia. Free Radical Research, 2002, 36, 467-469.	1.5	81
81	Respiratory-chain diseases related to complex III deficiency. Biochimica Et Biophysica Acta - Molecular Cell Research, 2009, 1793, 181-185.	1.9	81
82	Deficiency in complex II of the respiratory chain, presenting as a leukodystrophy in two sisters with leigh syndrome. Brain and Development, 1992, 14, 404-408.	0.6	78
83	A High Rate (20%â€“30%) of Parental Consanguinity in Cytochrome-Oxidase Deficiency. American Journal of Human Genetics, 1998, 63, 428-435.	2.6	77
84	A novel mutation in the dihydrolipoamide dehydrogenase E3 subunit gene (DLD) resulting in an atypical form of Î±-ketoglutarate dehydrogenase deficiency. Human Mutation, 2005, 25, 323-324.	1.1	76
85	The Variability of the Harlequin Mouse Phenotype Resembles that of Human Mitochondrial-Complex I-Deficiency Syndromes. PLoS ONE, 2008, 3, e3208.	1.1	74
86	Real-time flow cytometry analysis of permeability transition in isolated mitochondria. Experimental Cell Research, 2004, 294, 106-117.	1.2	69
87	A Simple and Accurate Spectrophotometric Assay for Phosphoenolpyruvate Carboxylase Activity. Plant Physiology, 1988, 86, 325-328.	2.3	68
88	Cytochrome c oxidase assay in minute amounts of human skeletal muscle using single wavelength spectrophotometers. Journal of Neuroscience Methods, 1998, 80, 107-111.	1.3	67
89	Unsuspected task for an old team: Succinate, fumarate and other Krebs cycle acids in metabolic remodeling. Biochimica Et Biophysica Acta - Bioenergetics, 2014, 1837, 1330-1337.	0.5	66
90	The measurement of the rotenone-sensitive NADH cytochrome c reductase activity in mitochondria isolated from minute amount of human skeletal muscle. Biochemical and Biophysical Research Communications, 1990, 173, 26-33.	1.0	63

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91	Quinone analogues regulate mitochondrial substrate competitive oxidation. Biochemical and Biophysical Research Communications, 2004, 316, 1138-1142.	1.0	62
92	Mutations in CYC1, Encoding Cytochrome c1 Subunit of Respiratory Chain Complex III, Cause Insulin-Responsive Hyperglycemia. American Journal of Human Genetics, 2013, 93, 384-389.	2.6	61
93	RXRÎ± overexpression in cardiomyocytes causes dilated cardiomyopathy but fails to rescue myocardial hypoplasia in RXRÎ±-null fetuses. Journal of Clinical Investigation, 2000, 105, 387-394.	3.9	61
94	Hepatic failure in disorders of oxidative phosphorylation with neonatal onset. Journal of Pediatrics, 1991, 119, 951-954.	0.9	60
95	Friedreich's ataxia: the vicious circle hypothesis revisited. BMC Medicine, 2011, 9, 112.	2.3	60
96	Mitochondrial dysfunction following perinatal exposure to nucleoside analogues. Aids, 2006, 20, 1685-1690.	1.0	59
97	Hereditary Paraganglioma/Pheochromocytoma and Inherited Succinate Dehydrogenase Deficiency. Hormone Research in Paediatrics, 2005, 63, 171-179.	0.8	57
98	Expression of alternative oxidase in Drosophila ameliorates diverse phenotypes due to cytochrome oxidase deficiency. Human Molecular Genetics, 2014, 23, 2078-2093.	1.4	57
99	Assay of mitochondrial respiratory chain complex I in human lymphocytes and cultured skin fibroblasts. Biochemical and Biophysical Research Communications, 2003, 301, 222-224.	1.0	56
100	Deferiprone targets aconitase: Implication for Friedreich's ataxia treatment. BMC Neurology, 2008, 8, 20.	0.8	55
101	Mitochondrial complex I deficiency of nuclear origin. Molecular Genetics and Metabolism, 2012, 105, 163-172.	0.5	55
102	Genotyping microsatellite DNA markers at putative disease loci in inbred/multiplex families with respiratory chain complexÂI deficiency allows rapid identification of a novel nonsense mutation (IVS1nt â^1) in the NDUFS4 gene in Leigh syndrome. Human Genetics, 2003, 112, 563-566.	1.8	54
103	In Vivo Detection of Succinate by Magnetic Resonance Spectroscopy as a Hallmark of SDHx Mutations in Paraganglioma. Clinical Cancer Research, 2016, 22, 1120-1129.	3.2	54
104	Refractory anaemia and mitochondrial cytopathy in childhood. British Journal of Haematology, 1994, 87, 381-385.	1.2	53
105	Supernumerary subunits NDUFA3, NDUFA5 and NDUFA12 are required for the formation of the extramembrane arm of human mitochondrial complex I. FEBS Letters, 2014, 588, 1832-1838.	1.3	53
106	Deletion of mitochondrial DNA in patient with chronic tubulointerstitial nephritis. Journal of Pediatrics, 1995, 126, 597-601.	0.9	52
107	Glutathione precursors replenish decreased glutathione pool in cystinotic cell lines. Biochemical and Biophysical Research Communications, 2004, 324, 231-235.	1.0	52
108	Fluxes of Nicotinamide Adenine Dinucleotides through Mitochondrial Membranes in Human Cultured Cells. Journal of Biological Chemistry, 1996, 271, 14785-14790.	1.6	50

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109	An improved spectrophotometric assay of pyruvate dehydrogenase in lactate dehydrogenase contaminated mitochondrial preparations from human skeletal muscle. Clinica Chimica Acta, 1995, 240, 129-136.	0.5	49
110	Letters to the Editor. Journal of Hepatology, 2000, 32, 364-365.	1.8	49
111	Engineering the alternative oxidase gene to better understand and counteract mitochondrial defects: state of the art and perspectives. British Journal of Pharmacology, 2014, 171, 2243-2249.	2.7	49
112	Glucose Modulates Respiratory Complex I Activity in Response to Acute Mitochondrial Dysfunction. Journal of Biological Chemistry, 2012, 287, 38729-38740.	1.6	46
113	Broad AOX expression in a genetically tractable mouse model does not disturb normal physiology. DMM Disease Models and Mechanisms, 2017, 10, 163-171.	1.2	46
114	QIL1 mutation causes MICOS disassembly and early onset fatal mitochondrial encephalopathy with liver disease. ELife, 2016, 5, .	2.8	46
115	Mitochondrial response to controlled nutrition in health and disease. Nutrition Reviews, 2011, 69, 65-75.	2.6	45
116	Animal models for respiratory chain disease. Trends in Molecular Medicine, 2001, 7, 578-581.	3.5	44
117	Respiratory chain deficiency presenting as congenital nephrotic syndrome. Pediatric Nephrology, 2005, 20, 465-469.	0.9	44
118	Evolutionarily conserved susceptibility of the mitochondrial respiratory chain to SDHI pesticides and its consequence on the impact of SDHIs on human cultured cells. PLoS ONE, 2019, 14, e0224132.	1.1	43
119	Cerebral white matter disease in children may be caused by mitochondrial respiratory chain deficiency. Journal of Pediatrics, 2000, 136, 209-214.	0.9	41
120	Respiratory chain alternative enzymes as tools to better understand and counteract respiratory chain deficiencies in human cells and animals. Physiologia Plantarum, 2009, 137, 362-370.	2.6	41
121	Use of Human Cancer Cell Lines Mitochondria to Explore the Mechanisms of BH3 Peptides and ABT-737-Induced Mitochondrial Membrane Permeabilization. PLoS ONE, 2010, 5, e9924.	1.1	41
122	Segregation of the G8993 mutant mitochondrial DNA through generations and embryonic tissues in a family at risk of leigh syndrome. Journal of Pediatrics, 1997, 131, 447-449.	0.9	40
123	Apoptosis-Inducing Factor Deficiency Induces Early Mitochondrial Degeneration in Brain Followed by Progressive Multifocal Neuropathology. Journal of Neuropathology and Experimental Neurology, 2007, 66, 838-847.	0.9	40
124	Mutations in the Dimer Interface of Dihydrolipoamide Dehydrogenase Promote Site-specific Oxidative Damages in Yeast and Human Cells. Journal of Biological Chemistry, 2011, 286, 40232-40245.	1.6	40
125	Idebenone treatment in Friedreich patients: One-year-long randomized placebo-controlled trial. Neurology, 2004, 62, 524-525.	1.5	38
126	CHCHD2 accumulates in distressed mitochondria and facilitates oligomerization of CHCHD10. Human Molecular Genetics, 2018, 27, 3881-3900.	1.4	38

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127	Presentation of six cases of StÃ¼ve-Wiedemann syndrome. Pediatric Radiology, 1998, 28, 776-780.	1.1	37
128	Respiratory Chain Defects May Present Only with Hypoglycemia. Journal of Clinical Endocrinology and Metabolism, 2005, 90, 3780-3785.	1.8	35
129	Synthesis and Preliminary Biological Evaluations of Ionic and Nonionic Amphiphilic Î±-Phenyl-N-tert-butylnitrone Derivatives. Journal of Medicinal Chemistry, 2003, 46, 5230-5237.	2.9	34
130	The gene responsible for Dyggve-Melchior-Clausen syndrome encodes a novel peripheral membrane protein dynamically associated with the Golgi apparatus. Human Molecular Genetics, 2009, 18, 440-453.	1.4	34
131	Longevity-relevant regulation of autophagy at the level of the acetylproteome. Autophagy, 2011, 7, 647-649.	4.3	34
132	Downregulation of apoptosis-inducing factor in Harlequin mice induces progressive and severe optic atrophy which is durably prevented by AAV2-AIF1 gene therapy. Brain, 2012, 135, 35-52.	3.7	34
133	Oxidative stress induces mitochondrial fragmentation in frataxin-deficient cells. Biochemical and Biophysical Research Communications, 2012, 418, 336-341.	1.0	34
134	From Nf1 to Sdhb knockout: Successes and failures in the quest for animal models of pheochromocytoma. Molecular and Cellular Endocrinology, 2016, 421, 40-48.	1.6	34
135	Respiratory chain defects: what do we know for sure about their consequences in vivo?. Biochimica Et Biophysica Acta - Bioenergetics, 2004, 1659, 172-177.	0.5	33
136	Interaction between Mitochondrial Cytochromes and Linoleic Acid Hydroperoxide. Plant Physiology, 1982, 69, 1308-1314.	2.3	32
137	Quinone analogs prevent enzymes targeted in Friedreich ataxia from ironâ€induced injury in vitro. BioFactors, 1999, 9, 247-251.	2.6	31
138	For debate: defective mitochondria, free radicals, cell death, aging-reality or myth-ochondria?. Mechanisms of Ageing and Development, 2000, 114, 201-206.	2.2	31
139	The alternative oxidase, a tool for compensating cytochrome c oxidase deficiency in human cells. Physiologia Plantarum, 2009, 137, 427-434.	2.6	31
140	Genetic background influences mitochondrial function: modeling mitochondrial disease for therapeutic development. Trends in Molecular Medicine, 2010, 16, 210-217.	3.5	31
141	KBPâ€“cytoskeleton interactions underlie developmental anomalies in Goldbergâ€“Shprintzen syndrome. Human Molecular Genetics, 2013, 22, 2387-2399.	1.4	31
142	Expression of the alternative oxidase mitigates beta-amyloid production and toxicity in model systems. Free Radical Biology and Medicine, 2016, 96, 57-66.	1.3	31
143	Coenzyme Q 10 Depletion is Comparatively Less Detrimental to Human Cultured Skin Fibroblasts than Respiratory Chain Complex Deficiencies. Free Radical Research, 2002, 36, 375-379.	1.5	30
144	Cytochrome c oxidase deficiency presenting as recurrent neonatal myoglobinuria. Neuromuscular Disorders, 1995, 5, 285-289.	0.3	29

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145	Cells Lacking the Fumarase Tumor Suppressor Are Protected from Apoptosis through a Hypoxia-Inducible Factor-Independent, AMPK-Dependent Mechanism. Molecular and Cellular Biology, 2012, 32, 3081-3094.	1.1	29
146	Mouse Studies to Shape Clinical Trials for Mitochondrial Diseases: High Fat Diet in Harlequin Mice. PLoS ONE, 2011, 6, e28823.	1.1	28
147	Translocator Protein-Mediated Stabilization of Mitochondrial Architecture during Inflammation Stress in Colonic Cells. PLoS ONE, 2016, 11, e0152919.	1.1	28
148	Kinetic Studies of the Form of Substrate Bound by Phosphoenolpyruvate Carboxylase. Plant Physiology, 1988, 88, 976-979.	2.3	27
149	Revisiting Pitfalls, Problems and Tentative Solutions for Assaying [General Articles] Mitochondrial Respiratory Chain Complex III in Human Samples. Current Medicinal Chemistry, 2004, 11, 233-239.	1.2	27
150	Mitochondrial complex I deficiency of nuclear origin. Molecular Genetics and Metabolism, 2012, 105, 173-179.	0.5	27
151	Mitochondria and diabetes mellitus: untangling a conflictive relationship?. Journal of Inherited Metabolic Disease, 2009, 32, 684-698.	1.7	26
152	Genetic and biochemical intricacy shapes mitochondrial cytopathies. Neurobiology of Disease, 2016, 92, 55-63.	2.1	26
153	Malate Metabolism in Leaf Mitochondria from the Crassulacean Acid Metabolism Plant KalanchoÃ« blossfeldiana Poelln. Plant Physiology, 1986, 81, 1039-1043.	2.3	25
154	Expression of respiratory chain deficiencies in human cultured cells. Neuromuscular Disorders, 1993, 3, 605-608.	0.3	25
155	The use of antioxidants in Friedreichâ€™s ataxia treatment. Expert Opinion on Investigational Drugs, 2003, 12, 569-575.	1.9	25
156	Cis-silencing of PIP5K1B evidenced in Friedreich's ataxia patient cells results in cytoskeleton anomalies. Human Molecular Genetics, 2013, 22, 2894-2904.	1.4	25
157	Effect of Lon protease knockdown on mitochondrial function in HeLa cells. Biochimie, 2014, 100, 38-47.	1.3	24
158	Oxidative interactions between fatty acid peroxy radicals and quinones: Possible involvement in cyanide-resistant electron transport in plant mitochondria. Archives of Biochemistry and Biophysics, 1983, 225, 630-639.	1.4	23
159	A kinetic study of the effects of phosphate and organic phosphates on the activity of phosphoenolpyruvate carboxylase from Crassula argentea. Archives of Biochemistry and Biophysics, 1989, 271, 84-97.	1.4	23
160	Cell Metabolic Alterations due to Mcph1 Mutation in Microcephaly. Cell Reports, 2020, 31, 107506.	2.9	23
161	The Consequences of a Mild Respiratory Chain Deficiency on Substrate Competitive Oxidation in Human Mitochondria. Biochemical and Biophysical Research Communications, 1997, 236, 643-646.	1.0	22
162	Malic enzyme activity and cyanide-insensitive electron transport in plant mitochondria. Biochemical and Biophysical Research Communications, 1979, 88, 1125-1131.	1.0	21

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163	Another observation with VATER association and a complex IV respiratory chain deficiency. European Journal of Medical Genetics, 2006, 49, 71-77.	0.7	21
164	Alternative respiratory chain enzymes: Therapeutic potential and possible pitfalls. Biochimica Et Biophysica Acta - Molecular Basis of Disease, 2019, 1865, 854-866.	1.8	21
165	Mitochondrial Respiratory Chain Dysfunction Caused by Coenzyme Q Deficiency. Methods in Enzymology, 2004, 382, 81-88.	0.4	20
166	A cancerâ€predisposing â€œhot spotâ€•mutation of the fumarase gene creates a dominant negative protein. International Journal of Cancer, 2008, 122, 947-951.	2.3	20
167	Therapies in inborn errors of oxidative metabolism. Trends in Endocrinology and Metabolism, 2012, 23, 488-495.	3.1	19
168	Pitfalls in Monitoring Mitochondrial Temperature Using Charged Thermosensitive Fluorophores. Chemosensors, 2020, 8, 124.	1.8	19
169	No mitochondrial cytochrome oxidase (COX) gene mutations in 18 cases of COX deficiency. Human Genetics, 1997, 101, 247.	1.8	18
170	Iron overload and mitochondrial diseases. Lancet, The, 1998, 351, 1286-1287.	6.3	17
171	Human cultured skin fibroblasts survive profound inherited ubiquinone depletion. Free Radical Research, 2001, 35, 11-21.	1.5	17
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