Nils-GÃ¶ran Larsson

List of Publications by Year
in descending order

184 papers	26,305 citations	6254 80 h-index	6471 157 g-index
188 all docs	188 docs citations	188 times ranked	23204 citing authors

#	Article	IF	Citations
1	Premature ageing in mice expressing defective mitochondrial DNA polymerase. Nature, 2004, 429, 417-423.	27.8	2,318
2	Mitochondrial transcription factor A is necessary for mtDNA maintance and embryogenesis in mice. Nature Genetics, 1998, 18, 231-236.	21.4	1,377
3	The role of mitochondria in aging. Journal of Clinical Investigation, 2013, 123, 951-957.	8.2	823
4	Mitochondrial transcription factor A regulates mtDNA copy number in mammals. Human Molecular Genetics, 2004, 13, 935-944.	2.9	730
5	Unraveling the Biological Roles of Reactive Oxygen Species. Cell Metabolism, 2011, 13, 361-366.	16.2	661
6	DNA Replication and Transcription in Mammalian Mitochondria. Annual Review of Biochemistry, 2007, 76, 679-699.	11.1	567
7	Mitochondrial transcription factors B1 and B2 activate transcription of human mtDNA. Nature Genetics, 2002, 31, 289-294.	21.4	535
8	Progressive parkinsonism in mice with respiratory-chain-deficient dopamine neurons. Proceedings of the National Academy of Sciences of the United States of America, 2007, 104, 1325-1330.	7.1	516
9	Maintenance and Expression of Mammalian Mitochondrial DNA. Annual Review of Biochemistry, 2016, 85, 133-160.	11.1	507
10	Somatic mtDNA mutations cause aging phenotypes without affecting reactive oxygen species production. Proceedings of the National Academy of Sciences of the United States of America, 2005, 102, 17993-17998.	7.1	491
11	Mammalian Mitochondria and Aging: An Update. Cell Metabolism, 2017, 25, 57-71.	16.2	463
12	Somatic Mitochondrial DNA Mutations in Mammalian Aging. Annual Review of Biochemistry, 2010, 79, 683-706.	11.1	456
13	MOLECULAR GENETIC ASPECTS OF HUMAN MITOCHONDRIAL DISORDERS. Annual Review of Genetics, 1995, 29, 151-178.	7.6	455
14	Super-resolution microscopy reveals that mammalian mitochondrial nucleoids have a uniform size and frequently contain a single copy of mtDNA. Proceedings of the National Academy of Sciences of the United States of America, 2011, 108, 13534-13539.	7.1	450
15	Strong Purifying Selection in Transmission of Mammalian Mitochondrial DNA. PLoS Biology, 2008, 6, e10.	5.6	425
16	Impaired insulin secretion and Î²-cell loss in tissue-specific knockout mice with mitochondrial diabetes. Nature Genetics, 2000, 26, 336-340.	21.4	417
17	Dilated cardiomyopathy and atrioventricular conduction blocks induced by heart-specific inactivation of mitochondrial DNA gene expression. Nature Genetics, 1999, 21, 133-137.	21.4	393
18	Proteolytic Processing of OPA1 Links Mitochondrial Dysfunction to Alterations in Mitochondrial Morphology. Journal of Biological Chemistry, 2006, 281, 37972-37979.	3.4	382

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19	Nurr1 Is Required for Maintenance of Maturing and Adult Midbrain Dopamine Neurons. Journal of Neuroscience, 2009, 29, 15923-15932.	3.6	320
20	Progressive Increase of the Mutated Mitochondrial DNA Fraction in Kearns-Sayre Syndrome. Pediatric Research, 1990, 28, 131-136.	2.3	308
21	Mitochondrial dysfunction as a cause of ageing. Journal of Internal Medicine, 2008, 263, 167-178.	6.0	292
22	The Enigma of the Respiratory Chain Supercomplex. Cell Metabolism, 2017, 25, 765-776.	16.2	279
23	Cross-strand binding of TFAM to a single mtDNA molecule forms the mitochondrial nucleoid. Proceedings of the National Academy of Sciences of the United States of America, 2015, 112, 11288-11293.	7.1	266
24	Methylation of 12S rRNA Is Necessary for In Vivo Stability of the Small Subunit of the Mammalian Mitochondrial Ribosome. Cell Metabolism, 2009, 9, 386-397.	16.2	264
25	LRPPRC is necessary for polyadenylation and coordination of translation of mitochondrial mRNAs. EMBO Journal, 2012, 31, 443-456.	7.8	264
26	Increased mitochondrial mass in mitochondrial myopathy mice. Proceedings of the National Academy of Sciences of the United States of America, 2002, 99, 15066-15071.	7.1	262
27	Impaired mitochondrial transport and Parkin-independent degeneration of respiratory chain-deficient dopamine neurons in vivo. Proceedings of the National Academy of Sciences of the United States of America, 2011, 108, 12937-12942.	7.1	258
28	Mitochondrial DNA mutations in disease and aging. Journal of Cell Biology, 2011, 193, 809-818.	5.2	242
29	Increased in vivo apoptosis in cells lacking mitochondrial DNA gene expression. Proceedings of the National Academy of Sciences of the United States of America, 2001, 98, 4038-4043.	7.1	234
30	The role of mitochondrial <scp>DNA</scp> mutations and free radicals in disease and ageing. Journal of Internal Medicine, 2013, 273, 529-543.	6.0	232
31	NSUN4 Is a Dual Function Mitochondrial Protein Required for Both Methylation of 12S rRNA and Coordination of Mitoribosomal Assembly. PLoS Genetics, 2014, 10, e1004110.	3.5	232
32	Germline mitochondrial DNA mutations aggravate ageing and can impair brain development. Nature, 2013, 501, 412-415.	27.8	231
33	MTERF4 Regulates Translation by Targeting the Methyltransferase NSUN4 to the Mammalian Mitochondrial Ribosome. Cell Metabolism, 2011, 13, 527-539.	16.2	221
34	Mitochondrial DNA copy number in human disease: the more the better?. FEBS Letters, 2021, 595, 976-1002.	2.8	219
35	High brain lactate is a hallmark of aging and caused by a shift in the lactate dehydrogenase A/B ratio. Proceedings of the National Academy of Sciences of the United States of America, 2010, 107, 20087-20092.	7.1	218
36	Somatic Progenitor Cell Vulnerability to Mitochondrial DNA Mutagenesis Underlies Progeroid Phenotypes in Polg Mutator Mice. Cell Metabolism, 2012, 15, 100-109.	16.2	213

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37	MTERF3 Is a Negative Regulator of Mammalian mtDNA Transcription. Cell, 2007, 130, 273-285.	28.9	209
38	Adipose-Specific Deletion of TFAM Increases Mitochondrial Oxidation and Protects Mice against Obesity and Insulin Resistance. Cell Metabolism, 2012, 16, 765-776.	16.2	206
39	Random Point Mutations with Major Effects on Protein-Coding Genes Are the Driving Force behind Premature Aging in mtDNA Mutator Mice. Cell Metabolism, 2009, 10, 131-138.	16.2	200
40	A switch in metabolism precedes increased mitochondrial biogenesis in respiratory chain-deficient mouse hearts. Proceedings of the National Academy of Sciences of the United States of America, 2004, 101, 3136-3141.	7.1	198
41	MitoTALEN reduces mutant mtDNA load and restores tRNAAla levels in a mouse model of heteroplasmic mtDNA mutation. Nature Medicine, 2018, 24, 1696-1700.	30.7	187
42	Mitofusin 2 is required to maintain mitochondrial coenzyme Q levels. Journal of Cell Biology, 2015, 208, 429-442.	5.2	180
43	Genetic modification of survival in tissue-specific knockout mice with mitochondrial cardiomyopathy. Proceedings of the National Academy of Sciences of the United States of America, 2000, 97, 3467-3472.	7.1	176
44	Making Proteins in the Powerhouse. Cell Metabolism, 2014, 20, 226-240.	16.2	170
45	Transcriptomic and proteomic landscape of mitochondrial dysfunction reveals secondary coenzyme Q deficiency in mammals. ELife, 2017, 6, .	6.0	169
46	Ultra-Deep Sequencing of Mouse Mitochondrial DNA: Mutational Patterns and Their Origins. PLoS Genetics, 2011, 7, e1002028.	3.5	162
47	Mammalian transcription factor A is a core component of the mitochondrial transcription machinery. Proceedings of the National Academy of Sciences of the United States of America, 2012, 109, 16510-16515.	7.1	156
48	Leber's hereditary optic neuropathy and complex I deficiency in muscle. Annals of Neurology, 1991, 30, 701-708.	5.3	155
49	Purifying selection of mtDNA and its implications for understanding evolution and mitochondrial disease. Nature Reviews Genetics, 2008, 9, 657-662.	16.3	155
50	Late-Onset Corticohippocampal Neurodepletion Attributable to Catastrophic Failure of Oxidative Phosphorylation in MILON Mice. Journal of Neuroscience, 2001, 21, 8082-8090.	3.6	151
51	The mitochondrial RNA polymerase contributes critically to promoter specificity in mammalian cells. EMBO Journal, 2004, 23, 4606-4614.	7.8	151
52	Mitofusin 2 is necessary for striatal axonal projections of midbrain dopamine neurons. Human Molecular Genetics, 2012, 21, 4827-4835.	2.9	149
53	A single mouse gene encodes the mitochondrial transcription factor A and a testisâ€“specific nuclear HMG-box protein. Nature Genetics, 1996, 13, 296-302.	21.4	145
54	Adipose tissue mitochondrial dysfunction triggers a lipodystrophic syndrome with insulin resistance, hepatosteatosis, and cardiovascular complications. FASEB Journal, 2014, 28, 4408-4419.	0.5	136

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55	Respiratory chain dysfunction in skeletal muscle does not cause insulin resistance. Biochemical and Biophysical Research Communications, 2006, 350, 202-207.	2.1	134
56	TWINKLE is an essential mitochondrial helicase required for synthesis of nascent D-loop strands and complete mtDNA replication. Human Molecular Genetics, 2013, 22, 1983-1993.	2.9	132
57	Mitochondrial encephalomyopathies in childhood. I. Biochemical and morphologic investigations. Journal of Pediatrics, 1991, 119, 242-250.	1.8	128
58	Variation in germline mtDNA heteroplasmy is determined prenatally but modified during subsequent transmission. Nature Genetics, 2012, 44, 1282-1285.	21.4	128
59	Mitochondrial DNA deletions in inclusion body myositis. Brain, 1993, 116, 325-336.	7.6	116
60	A family of putative transcription termination factors shared amongst metazoans and plants. Current Genetics, 2005, 48, 265-269.	1.7	116
61	Reactive oxygen species and fatigueâ€induced prolonged lowâ€frequency force depression in skeletal muscle fibres of rats, mice and SOD2 overexpressing mice. Journal of Physiology, 2008, 586, 175-184.	2.9	116
62	Hierarchical RNA Processing Is Required for Mitochondrial Ribosome Assembly. Cell Reports, 2016, 16, 1874-1890.	6.4	116
63	Mitochondrial fusion is required for regulation of mitochondrial DNA replication. PLoS Genetics, 2019, 15, e1008085.	3.5	116
64	Small-molecule inhibitors of human mitochondrial DNA transcription. Nature, 2020, 588, 712-716.	27.8	115
65	Impaired nigrostriatal function precedes behavioral deficits in a genetic mitochondrial model of Parkinson's disease. FASEB Journal, 2011, 25, 1333-1344.	0.5	112
66	Changes of mitochondrial ultrastructure and function during ageing in mice and Drosophila. ELife, 2017, 6, .	6.0	108
67	mtDNA makes a U-turn for the mitochondrial nucleoid. Trends in Cell Biology, 2013, 23, 457-463.	7.9	106
68	Structure of the human MTERF4â€“NSUN4 protein complex that regulates mitochondrial ribosome biogenesis. Proceedings of the National Academy of Sciences of the United States of America, 2012, 109, 15253-15258.	7.1	105
69	Mitochondrial DNA Deletions in Muscle Fibers in Inclusion Body Myositis. Journal of Neuropathology and Experimental Neurology, 1995, 54, 581-587.	1.7	103
70	A Phenotype-Driven Approach to Generate Mouse Models with Pathogenic mtDNA Mutations Causing Mitochondrial Disease. Cell Reports, 2016, 16, 2980-2990.	6.4	102
71	Topoisomerase 3Î± Is Required for Decatenation and Segregation of Human mtDNA. Molecular Cell, 2018, 69, 9-23.e6.	9.7	102
72	SOD2 overexpression: enhanced mitochondrial tolerance but absence of effect on UCP activity. EMBO Journal, 2005, 24, 4061-4070.	7.8	98

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73	The transcription machinery in mammalian mitochondria. Biochimica Et Biophysica Acta - Bioenergetics, 2004, 1659, 148-152.	1.0	94
74	MTERF1 Binds mtDNA to Prevent Transcriptional Interference at the Light-Strand Promoter but Is Dispensable for rRNA Gene Transcription Regulation. Cell Metabolism, 2013, 17, 618-626.	16.2	93
75	COX7A2L Is a Mitochondrial Complex III Binding Protein that Stabilizes the III2+IV Supercomplex without Affecting Respirasome Formation. Cell Reports, 2016, 16, 2387-2398.	6.4	93
76	MitoPark mice mirror the slow progression of key symptoms and Lâ€DOPA response in Parkinson's disease. Genes, Brain and Behavior, 2010, 9, 173-181.	2.2	92
77	No recombination of mtDNA after heteroplasmy for 50 generations in the mouse maternal germline. Nucleic Acids Research, 2014, 42, 1111-1116.	14.5	92
78	Loss of LRPPRC causes ATP synthase deficiency. Human Molecular Genetics, 2014, 23, 2580-2592.	2.9	91
79	POLRMT regulates the switch between replication primer formation and gene expression of mammalian mtDNA. Science Advances, 2016, 2, e1600963.	10.3	91
80	Keeping mtDNA in Shape between Generations. PLoS Genetics, 2014, 10, e1004670.	3.5	90
81	The Respiratory Chain Supercomplex Organization Is Independent of COX7a2l Isoforms. Cell Metabolism, 2014, 20, 1069-1075.	16.2	90
82	AFG3L2 supports mitochondrial protein synthesis and Purkinje cell survival. Journal of Clinical Investigation, 2012, 122, 4048-4058.	8.2	90
83	Modulation of mtDNA copy number ameliorates the pathological consequences of a heteroplasmic mtDNA mutation in the mouse. Science Advances, 2019, 5, eaav9824.	10.3	86
84	Mitochondrial encephalomyopathies in childhood. II. Clinical manifestations and syndromes. Journal of Pediatrics, 1991, 119, 251-259.	1.8	85
85	MTERF3 Regulates Mitochondrial Ribosome Biogenesis in Invertebrates and Mammals. PLoS Genetics, 2013, 9, e1003178.	3.5	85
86	Increased Total mtDNA Copy Number Cures Male Infertility Despite Unaltered mtDNA Mutation Load. Cell Metabolism, 2017, 26, 429-436.e4.	16.2	84
87	A Common Variant in TFB1M Is Associated with Reduced Insulin Secretion and Increased Future Risk of Type 2 Diabetes. Cell Metabolism, 2011, 13, 80-91.	16.2	81
88	Understanding aging: revealing order out of chaos. Biochimica Et Biophysica Acta - Bioenergetics, 2004, 1658, 122-132.	1.0	80
89	SLIRP Regulates the Rate of Mitochondrial Protein Synthesis and Protects LRPPRC from Degradation. PLoS Genetics, 2015, 11, e1005423.	3.5	80
90	LRPPRC-mediated folding of the mitochondrial transcriptome. Nature Communications, 2017, 8, 1532.	12.8	80

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91	Down-Regulation of Mitochondrial Transcription Factor a During Spermatogenesis in Humans. Human Molecular Genetics, 1997, 6, 185-1991.	2.9	75
92	Loss of the RNA-binding protein TACO1 causes late-onset mitochondrial dysfunction in mice. Nature Communications, 2016, 7, 11884.	12.8	73
93	Mitochondrial medicine - recent advances. Journal of Internal Medicine, 1999, 246, 11-23.	6.0	70
94	MTERF2 is a nucleoid component in mammalian mitochondria. Biochimica Et Biophysica Acta - Bioenergetics, 2009, 1787, 296-302.	1.0	70
95	Altered dopamine metabolism and increased vulnerability to MPTP in mice with partial deficiency of mitochondrial complex I in dopamine neurons. Human Molecular Genetics, 2012, 21, 1078-1089.	2.9	69
96	Revolution in mitochondrial medicine. FEBS Letters, 1999, 455, 199-202.	2.8	66
97	Downregulation of Tfam and mtDNA copy number during mammalian spermatogenesis. Mammalian Genome, 2001, 12, 787-792.	2.2	66
98	Mitochondrial ATP-Synthase Deficiency in a Child with 3-Methylglutaconic Aciduria. Pediatric Research, 1992, 32, 731-736.	2.3	65
99	Increased mitochondrial Ca 2+ and decreased sarcoplasmic reticulum Ca 2+ in mitochondrial myopathy. Human Molecular Genetics, 2009, 18, 278-288.	2.9	64
100	Cellular pyrimidine imbalance triggers mitochondrial DNAâ€“dependent innate immunity. Nature Metabolism, 2021, 3, 636-650.	11.9	64
101	A human mitochondrial poly(A) polymerase mutation reveals the complexities of post-transcriptional mitochondrial gene expression. Human Molecular Genetics, 2014, 23, 6345-6355.	2.9	63
102	Mitochondrial Dysfunction in Mammalian Ageing. Novartis Foundation Symposium, 2007, 287, 197-213.	1.1	63
103	Mitochondrial DNA deletions and cytochrome c oxidase deficiency in muscle fibres. Journal of the Neurological Sciences, 1992, 110, 169-177.	0.6	61
104	Pathogenetic aspects of the A8344G mutation of mitochondrial DNA associated with MERRF syndrome and multiple symmetric lipomas. Muscle and Nerve, 1995, 18, S102-S106.	2.2	59
105	In vivo mutagenesis reveals that OriL is essential for mitochondrial DNA replication. EMBO Reports, 2012, 13, 1130-1137.	4.5	59
106	Base-excision repair deficiency alone or combined with increased oxidative stress does not increase mtDNA point mutations in mice. Nucleic Acids Research, 2018, 46, 6642-6669.	14.5	58
107	Mice lacking the mitochondrial exonuclease MGME1 accumulate mtDNA deletions without developing progeria. Nature Communications, 2018, 9, 1202.	12.8	57
108	The Bicoid Stability Factor Controls Polyadenylation and Expression of Specific Mitochondrial mRNAs in Drosophila melanogaster. PLoS Genetics, 2011, 7, e1002324.	3.5	55

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109	Bioenergetic roles of mitochondrial fusion. Biochimica Et Biophysica Acta - Bioenergetics, 2016, 1857, 1277-1283.	1.0	55
110	Mitochondrial myopathies. Acta Physiologica Scandinavica, 2001, 171, 385-393.	2.2	54
111	Secondary metabolic effects in complex I deficiency. Annals of Neurology, 2005, 58, 544-552.	5.3	51
112	MtDNA mutations are a common cause of severe disease phenotypes in children with Leigh syndrome. Biochimica Et Biophysica Acta - Bioenergetics, 2009, 1787, 484-490.	1.0	51
113	The Mitochondrial Electron Transport Chain Is Dispensable for Proliferation and Differentiation of Epidermal Progenitor Cells. Stem Cells, 2011, 29, 1459-1468.	3.2	51
114	Loss of TFB1M results in mitochondrial dysfunction that leads to impaired insulin secretion and diabetes. Human Molecular Genetics, 2014, 23, 5733-5749.	2.9	51
115	PTCD1 Is Required for 16S rRNA Maturation Complex Stability and Mitochondrial Ribosome Assembly. Cell Reports, 2018, 23, 127-142.	6.4	51
116	<scp>TEFM</scp> regulates both transcription elongation and <scp>RNA</scp> processing in mitochondria. EMBO Reports, 2019, 20, .	4.5	51
117	The amino terminal extension of mammalian mitochondrial RNA polymerase ensures promoter specific transcription initiation. Nucleic Acids Research, 2014, 42, 3638-3647.	14.5	50
118	LRPPRC is a mitochondrial matrix protein that is conserved in metazoans. Biochemical and Biophysical Research Communications, 2010, 398, 759-764.	2.1	49
119	Somatic mtDNA mutations cause progressive hearing loss in the mouse. Experimental Cell Research, 2007, 313, 3924-3934.	2.6	48
120	Complementation between polymerase- and exonuclease-deficient mitochondrial DNA polymerase mutants in genomically engineered flies. Nature Communications, 2015, 6, 8808.	12.8	48
121	Automatic sequencing of mitochondrial tRNA genes in patients with mitochondrial encephalomyopathy. Biochimica Et Biophysica Acta - Molecular Basis of Disease, 1994, 1226, 49-55.	3.8	47
122	SnapShot: Mitochondrial Nucleoid. Cell, 2018, 172, 388-388.e1.	28.9	47
123	Neuronal metabolic rewiring promotes resilience to neurodegeneration caused by mitochondrial dysfunction. Science Advances, 2020, 6, eaba8271.	10.3	47
124	Animal models for respiratory chain disease. Trends in Molecular Medicine, 2001, 7, 578-581.	6.7	44
125	<scp>FBXL</scp> 4 deficiency increases mitochondrial removal by autophagy. EMBO Molecular Medicine, 2020, 12, e11659.	6.9	44
126	TFAM forces mtDNA to make a U-turn. Nature Structural and Molecular Biology, 2011, 18, 1179-1181.	8.2	43

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127	Age-associated mosaic respiratory chain deficiency causes trans-neuronal degeneration. Human Molecular Genetics, 2008, 17, 1418-1426.	2.9	41
128	Accurate mapping of mitochondrial DNA deletions and duplications using deep sequencing. PLoS Genetics, 2020, 16, e1009242.	3.5	41
129	Two novel mutations in thymidine kinase-2 cause early onset fatal encephalomyopathy and severe mtDNA depletion. Neuromuscular Disorders, 2010, 20, 198-203.	0.6	39
130	SLIRP stabilizes LRPPRC via an RRMâ€“PPR protein interface. Nucleic Acids Research, 2016, 44, 6868-6882.	14.5	39
131	High levels of TFAM repress mammalian mitochondrial DNA transcription in vivo. Life Science Alliance, 2021, 4, e202101034.	2.8	38
132	Sensory Ataxic Neuropathy in Golden Retriever Dogs Is Caused by a Deletion in the Mitochondrial tRNATyr Gene. PLoS Genetics, 2009, 5, e1000499.	3.5	37
133	Regulation of mitochondrial DNA copy number during spermatogenesis. Human Reproduction, 2000, 15, 86-91.	0.9	36
134	The mitochondrial single-stranded DNA binding protein is essential for initiation of mtDNA replication. Science Advances, 2021, 7, .	10.3	36
135	De novo mutation in the mitochondrial ATP synthase subunit 6 gene (T8993G) with rapid segregation resulting in Leigh syndrome in the offspring. Human Genetics, 1995, 96, 290-4.	3.8	35
136	Maintenance of respiratory chain function in mouse hearts with severely impaired mtDNA transcription. Nucleic Acids Research, 2010, 38, 6577-6588.	14.5	35
137	Defects in Î²-Cell Ca2+ Dynamics in Age-Induced Diabetes. Diabetes, 2014, 63, 4100-4114.	0.6	35
138	POLRMT does not transcribe nuclear genes. Nature, 2014, 514, E7-E11.	27.8	35
139	Characterization of the mouse genes for mitochondrial transcription factors B1 and B2. Mammalian Genome, 2003, 14, 1-6.	2.2	34
140	Similar patterns of clonally expanded somatic mtDNA mutations in the colon of heterozygous mtDNA mutator mice and ageing humans. Mechanisms of Ageing and Development, 2014, 139, 22-30.	4.6	33
141	Mutations of mitochondrial DNA are not major contributors to aging of fruit flies. Proceedings of the National Academy of Sciences of the United States of America, 2018, 115, E9620-E9629.	7.1	32
142	Atypical presentation of multisystem disorders in two girls with mitochondrial DNA deletions. European Journal of Pediatrics, 1995, 154, 35-42.	2.7	30
143	Drosophila melanogaster LRPPRC2 is involved in coordination of mitochondrial translation. Nucleic Acids Research, 2014, 42, 13920-13938.	14.5	29
144	The Leucine-rich Pentatricopeptide Repeat-containing Protein (LRPPRC) Does Not Activate Transcription in Mammalian Mitochondria. Journal of Biological Chemistry, 2013, 288, 15510-15519.	3.4	27

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145	Manipulation of mitochondrial DNA gene expression in the mouse. Biochimica Et Biophysica Acta - Bioenergetics, 2002, 1555, 106-110.	1.0	25
146	Sequence-specific stalling of DNA polymerase Î³ and the effects of mutations causing progressive ophthalmoplegiaâ€. Human Molecular Genetics, 2011, 20, 1212-1223.	2.9	24
147	MitoRibo-Tag Mice Provide a Tool for InÂVivo Studies of Mitoribosome Composition. Cell Reports, 2019, 29, 1728-1738.e9.	6.4	24
148	Abnormal Ca2+ release and catecholamine-induced arrhythmias in mitochondrial cardiomyopathy. Human Molecular Genetics, 2005, 14, 1069-1076.	2.9	22
149	Dinucleotide Degradation by REXO2 Maintains Promoter Specificity in Mammalian Mitochondria. Molecular Cell, 2019, 76, 784-796.e6.	9.7	22
150	Fatal mitochondrial myopathy, lactic acidosis, and complex I deficiency associated with a heteroplasmic Aâ†'G mutation at position 3251 in the mitochondrial tRNALeu(UUR) gene. Human Genetics, 1996, 97, 269-273.	3.8	20
151	Tracing the Trail of Protons through Complex I of the Mitochondrial Respiratory Chain. PLoS Biology, 2011, 9, e1001129.	5.6	19
152	Complex genetic counselling and prenatal analysis in a woman with external ophthalmoplegia and deleted mtDNA. , 2000, 20, 426-431.		18
153	Mitochondrial diseases. Best Practice and Research in Clinical Obstetrics and Gynaecology, 2002, 16, 715-728.	2.8	17
154	The UbL protein UBTD1 stably interacts with the UBE2D family of E2 ubiquitin conjugating enzymes. Biochemical and Biophysical Research Communications, 2014, 443, 7-12.	2.1	17
155	Metabolic resistance to the inhibition of mitochondrial transcription revealed by CRISPRâ€Cas9 screen. EMBO Reports, 2022, 23, e53054.	4.5	16
156	Leber hereditary optic neuropathy: A nuclear solution of a mitochondrial problem. Annals of Neurology, 2002, 52, 529-530.	5.3	15
157	Mic10 Oligomerization Pinches off Mitochondrial Cristae. Cell Metabolism, 2015, 21, 660-661.	16.2	14
158	Stable transfection of a human lymphoma line by sub-genomic fragments of Epstein-Barr virus DNA to measure humoral and cellular immunity to the corresponding proteins. International Journal of Cancer, 1987, 40, 389-395.	5.1	13
159	Multiple short direct repeats associated with single mtDNA deletions. Biochimica Et Biophysica Acta - Molecular Basis of Disease, 1992, 1139, 311-314.	3.8	13
160	Breeding and Genotyping of Tfam Conditional Knockout Mice. , 2002, 197, 391-400.		13
161	Is Energy Deficiency Good in Moderation?. Cell, 2007, 131, 448-450.	28.9	13
162	Point Mutations Are Causing Progeroid Phenotypes in the mtDNA Mutator Mouse. Cell Metabolism, 2010, 11, 1.	16.2	12

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163	Overexpression of the mitochondrial methyltransferase TFB1M in the mouse does not impact mitoribosomal methylation status or hearing. Human Molecular Genetics, 2015, 24, 7286-7294.	2.9	12
164	Enhanced Cardiomyocyte Ca²⁺ Cycling Precedes Terminal AV-Block in Mitochondrial Cardiomyopathy Mterf3 KO Mice. Antioxidants and Redox Signaling, 2011, 15, 2455-2464.	5.4	11
165	Tissue-Specific Knockout Model for Study of Mitochondrial DNA Mutation Disorders. Methods in Enzymology, 2002, 353, 409-421.	1.0	9
166	Mice lacking the mitochondrial exonuclease MGME1 develop inflammatory kidney disease with glomerular dysfunction. PLoS Genetics, 2022, 18, e1010190.	3.5	9
167	Mitochondrial dysfunction in adult midbrain dopamine neurons triggers an early immune response. PLoS Genetics, 2021, 17, e1009822.	3.5	8
168	Complex I: A Complex Gateway to the Powerhouse. Cell Metabolism, 2008, 7, 278-279.	16.2	7
169	Proofreading deficiency in mitochondrial DNA polymerase does not affect total dNTP pools in mouse embryos. Nature Metabolism, 2020, 2, 673-675.	11.9	7
170	Mitochondria in human disease. Journal of Internal Medicine, 2020, 287, 589-591.	6.0	7
171	Structure Casts Light on mtDNA Replication. Cell, 2009, 139, 231-233.	28.9	6
172	Response: Point Mutations Are Causing Progeroid Phenotypes in the mtDNA Mutator Mouse. Cell Metabolism, 2010, 11, 93.	16.2	5
173	Complete Deletion of a POLG1 Allele in a Patient with Alpers Syndrome. JIMD Reports, 2011, 4, 67-73.	1.5	5
174	An Adaptable High-Throughput Technology Enabling the Identification of Specific Transcription Modulators. SLAS Discovery, 2017, 22, 378-386.	2.7	5
175	USE OF PRIMARY CULTURES AND CONTINUOUS CELL LINES TO STUDY EFFECTS ON ASTROCYTIC REGULATORY FUNCTIONS. Clinical and Experimental Pharmacology and Physiology, 1995, 22, 284-287.	1.9	3
176	MTERF1 Gives mtDNA an Unusual Twist. Cell Metabolism, 2010, 12, 3-4.	16.2	3
177	MitoTALEN reduces mutant mtDNA load and restores tRNAAla levels in a mouse model of heteroplasmic mtDNA mutation. , 0, .		1
178	Defective assembly of the respiratory chain. Acta Paediatrica, International Journal of Paediatrics, 2004, 93, 1268-1270.	1.5	0
179	Introduction: Biology of ageing. Journal of Internal Medicine, 2008, 263, 114-116.	6.0	0
180	Introduction: Euromit VII â€“ summarizing 50â€ƒyears of research on mitochondrial disease. Journal of Internal Medicine, 2009, 265, 170-173.	6.0	0

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181	Accurate mapping of mitochondrial DNA deletions and duplications using deep sequencing. , 2020, 16, e1009242.		0
182	Accurate mapping of mitochondrial DNA deletions and duplications using deep sequencing. , 2020, 16, e1009242.		0
183	Accurate mapping of mitochondrial DNA deletions and duplications using deep sequencing. , 2020, 16, e1009242.		0
184	Accurate mapping of mitochondrial DNA deletions and duplications using deep sequencing. , 2020, 16, e1009242.		0