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	Metabolic resistance to the inhibition of mitochondrial transcription revealed by CRISPRâ€Cas9 screen. EMBO Reports, 2022, 23, e53054.	4.5	16
2	Mice lacking the mitochondrial exonuclease MGME1 develop inflammatory kidney disease with glomerular dysfunction. PLoS Genetics, 2022, 18, e1010190.	3.5	9
3	Mitochondrial DNA copy number in human disease: the more the better?. FEBS Letters, 2021, 595, 976-1002.	2.8	219
4	Cellular pyrimidine imbalance triggers mitochondrial DNAâ€“dependent innate immunity. Nature Metabolism, 2021, 3, 636-650.	11.9	64
5	The mitochondrial single-stranded DNA binding protein is essential for initiation of mtDNA replication. Science Advances, 2021, 7, .	10.3	36
6	High levels of TFAM repress mammalian mitochondrial DNA transcription in vivo. Life Science Alliance, 2021, 4, e202101034.	2.8	38
7	Mitochondrial dysfunction in adult midbrain dopamine neurons triggers an early immune response. PLoS Genetics, 2021, 17, e1009822.	3.5	8
8	Proofreading deficiency in mitochondrial DNA polymerase does not affect total dNTP pools in mouse embryos. Nature Metabolism, 2020, 2, 673-675.	11.9	7
9	<scp>FBXL</scp> 4 deficiency increases mitochondrial removal by autophagy. EMBO Molecular Medicine, 2020, 12, e11659.	6.9	44
10	Neuronal metabolic rewiring promotes resilience to neurodegeneration caused by mitochondrial dysfunction. Science Advances, 2020, 6, eaba8271.	10.3	47
11	Small-molecule inhibitors of human mitochondrial DNA transcription. Nature, 2020, 588, 712-716.	27.8	115
12	Mitochondria in human disease. Journal of Internal Medicine, 2020, 287, 589-591.	6.0	7
13	Accurate mapping of mitochondrial DNA deletions and duplications using deep sequencing. PLoS Genetics, 2020, 16, e1009242.	3.5	41
14	Accurate mapping of mitochondrial DNA deletions and duplications using deep sequencing. , 2020, 16, e1009242.		0
15	Accurate mapping of mitochondrial DNA deletions and duplications using deep sequencing. , 2020, 16, e1009242.		0
16	Accurate mapping of mitochondrial DNA deletions and duplications using deep sequencing. , 2020, 16, e1009242.		0
17	Accurate mapping of mitochondrial DNA deletions and duplications using deep sequencing. , 2020, 16, e1009242.		0
18	MitoRibo-Tag Mice Provide a Tool for InÂVivo Studies of Mitoribosome Composition. Cell Reports, 2019, 29, 1728-1738.e9.	6.4	24

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19	Dinucleotide Degradation by REXO2 Maintains Promoter Specificity in Mammalian Mitochondria. Molecular Cell, 2019, 76, 784-796.e6.	9.7	22
20	Mitochondrial fusion is required for regulation of mitochondrial DNA replication. PLoS Genetics, 2019, 15, e1008085.	3.5	116
21	<scp>TEFM</scp> regulates both transcription elongation and <scp>RNA</scp> processing in mitochondria. EMBO Reports, 2019, 20, .	4.5	51
22	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
23	PTCD1 Is Required for 16S rRNA Maturation Complex Stability and Mitochondrial Ribosome Assembly. Cell Reports, 2018, 23, 127-142.	6.4	51
24	SnapShot: Mitochondrial Nucleoid. Cell, 2018, 172, 388-388.e1.	28.9	47
25	Topoisomerase 3Î± Is Required for Decatenation and Segregation of Human mtDNA. Molecular Cell, 2018, 69, 9-23.e6.	9.7	102
26	Mice lacking the mitochondrial exonuclease MGME1 accumulate mtDNA deletions without developing progeria. Nature Communications, 2018, 9, 1202.	12.8	57
27	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
28	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
29	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
30	An Adaptable High-Throughput Technology Enabling the Identification of Specific Transcription Modulators. SLAS Discovery, 2017, 22, 378-386.	2.7	5
31	The Enigma of the Respiratory Chain Supercomplex. Cell Metabolism, 2017, 25, 765-776.	16.2	279
32	Increased Total mtDNA Copy Number Cures Male Infertility Despite Unaltered mtDNA Mutation Load. Cell Metabolism, 2017, 26, 429-436.e4.	16.2	84
33	LRPPRC-mediated folding of the mitochondrial transcriptome. Nature Communications, 2017, 8, 1532.	12.8	80
34	Mammalian Mitochondria and Aging: An Update. Cell Metabolism, 2017, 25, 57-71.	16.2	463
35	Changes of mitochondrial ultrastructure and function during ageing in mice and Drosophila. ELife, 2017, 6, .	6.0	108
36	Transcriptomic and proteomic landscape of mitochondrial dysfunction reveals secondary coenzyme Q deficiency in mammals. ELife, 2017, 6, .	6.0	169

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37	Bioenergetic roles of mitochondrial fusion. Biochimica Et Biophysica Acta - Bioenergetics, 2016, 1857, 1277-1283.	1.0	55
38	A Phenotype-Driven Approach to Generate Mouse Models with Pathogenic mtDNA Mutations Causing Mitochondrial Disease. Cell Reports, 2016, 16, 2980-2990.	6.4	102
39	Hierarchical RNA Processing Is Required for Mitochondrial Ribosome Assembly. Cell Reports, 2016, 16, 1874-1890.	6.4	116
40	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
41	Loss of the RNA-binding protein TACO1 causes late-onset mitochondrial dysfunction in mice. Nature Communications, 2016, 7, 11884.	12.8	73
42	POLRMT regulates the switch between replication primer formation and gene expression of mammalian mtDNA. Science Advances, 2016, 2, e1600963.	10.3	91
43	SLIRP stabilizes LRPPRC via an RRMâ€“PPR protein interface. Nucleic Acids Research, 2016, 44, 6868-6882.	14.5	39
44	Maintenance and Expression of Mammalian Mitochondrial DNA. Annual Review of Biochemistry, 2016, 85, 133-160.	11.1	507
45	SLIRP Regulates the Rate of Mitochondrial Protein Synthesis and Protects LRPPRC from Degradation. PLoS Genetics, 2015, 11, e1005423.	3.5	80
46	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
47	Complementation between polymerase- and exonuclease-deficient mitochondrial DNA polymerase mutants in genomically engineered flies. Nature Communications, 2015, 6, 8808.	12.8	48
48	Mitofusin 2 is required to maintain mitochondrial coenzyme Q levels. Journal of Cell Biology, 2015, 208, 429-442.	5.2	180
49	Mic10 Oligomerization Pinches off Mitochondrial Cristae. Cell Metabolism, 2015, 21, 660-661.	16.2	14
50	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
51	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
52	Loss of LRPPRC causes ATP synthase deficiency. Human Molecular Genetics, 2014, 23, 2580-2592.	2.9	91
53	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
54	Keeping mtDNA in Shape between Generations. PLoS Genetics, 2014, 10, e1004670.	3.5	90

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55	The Respiratory Chain Supercomplex Organization Is Independent of COX7a2l Isoforms. Cell Metabolism, 2014, 20, 1069-1075.	16.2	90
56	Drosophila melanogaster LRPPRC2 is involved in coordination of mitochondrial translation. Nucleic Acids Research, 2014, 42, 13920-13938.	14.5	29
57	The amino terminal extension of mammalian mitochondrial RNA polymerase ensures promoter specific transcription initiation. Nucleic Acids Research, 2014, 42, 3638-3647.	14.5	50
58	No recombination of mtDNA after heteroplasmy for 50 generations in the mouse maternal germline. Nucleic Acids Research, 2014, 42, 1111-1116.	14.5	92
59	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
60	Defects in Î²-Cell Ca2+ Dynamics in Age-Induced Diabetes. Diabetes, 2014, 63, 4100-4114.	0.6	35
61	POLRMT does not transcribe nuclear genes. Nature, 2014, 514, E7-E11.	27.8	35
62	Adipose tissue mitochondrial dysfunction triggers a lipodystrophic syndrome with insulin resistance, hepatosteatosis, and cardiovascular complications. FASEB Journal, 2014, 28, 4408-4419.	0.5	136
63	Making Proteins in the Powerhouse. Cell Metabolism, 2014, 20, 226-240.	16.2	170
64	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
65	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
66	Germline mitochondrial DNA mutations aggravate ageing and can impair brain development. Nature, 2013, 501, 412-415.	27.8	231
67	mtDNA makes a U-turn for the mitochondrial nucleoid. Trends in Cell Biology, 2013, 23, 457-463.	7.9	106
68	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
69	The role of mitochondria in aging. Journal of Clinical Investigation, 2013, 123, 951-957.	8.2	823
70	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
71	MTERF3 Regulates Mitochondrial Ribosome Biogenesis in Invertebrates and Mammals. PLoS Genetics, 2013, 9, e1003178.	3.5	85
72	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

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73	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
74	LRPPRC is necessary for polyadenylation and coordination of translation of mitochondrial mRNAs. EMBO Journal, 2012, 31, 443-456.	7.8	264
75	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
76	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
77	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
78	Variation in germline mtDNA heteroplasmy is determined prenatally but modified during subsequent transmission. Nature Genetics, 2012, 44, 1282-1285.	21.4	128
79	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
80	Mitofusin 2 is necessary for striatal axonal projections of midbrain dopamine neurons. Human Molecular Genetics, 2012, 21, 4827-4835.	2.9	149
81	Somatic Progenitor Cell Vulnerability to Mitochondrial DNA Mutagenesis Underlies Progeroid Phenotypes in Polg Mutator Mice. Cell Metabolism, 2012, 15, 100-109.	16.2	213
82	In vivo mutagenesis reveals that OriL is essential for mitochondrial DNA replication. EMBO Reports, 2012, 13, 1130-1137.	4.5	59
83	AFG3L2 supports mitochondrial protein synthesis and Purkinje cell survival. Journal of Clinical Investigation, 2012, 122, 4048-4058.	8.2	90
84	TFAM forces mtDNA to make a U-turn. Nature Structural and Molecular Biology, 2011, 18, 1179-1181.	8.2	43
85	Mitochondrial DNA mutations in disease and aging. Journal of Cell Biology, 2011, 193, 809-818.	5.2	242
86	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
87	Unraveling the Biological Roles of Reactive Oxygen Species. Cell Metabolism, 2011, 13, 361-366.	16.2	661
88	MTERF4 Regulates Translation by Targeting the Methyltransferase NSUN4 to the Mammalian Mitochondrial Ribosome. Cell Metabolism, 2011, 13, 527-539.	16.2	221
89	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
90	The Mitochondrial Electron Transport Chain Is Dispensable for Proliferation and Differentiation of Epidermal Progenitor Cells. Stem Cells, 2011, 29, 1459-1468.	3.2	51

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91	Complete Deletion of a POLG1 Allele in a Patient with Alpers Syndrome. JIMD Reports, 2011, 4, 67-73.	1.5	5
92	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
93	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
94	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
95	Impaired nigrostriatal function precedes behavioral deficits in a genetic mitochondrial model of Parkinson's disease. FASEB Journal, 2011, 25, 1333-1344.	0.5	112
96	The Bicoid Stability Factor Controls Polyadenylation and Expression of Specific Mitochondrial mRNAs in Drosophila melanogaster. PLoS Genetics, 2011, 7, e1002324.	3.5	55
97	Tracing the Trail of Protons through Complex I of the Mitochondrial Respiratory Chain. PLoS Biology, 2011, 9, e1001129.	5.6	19
98	Ultra-Deep Sequencing of Mouse Mitochondrial DNA: Mutational Patterns and Their Origins. PLoS Genetics, 2011, 7, e1002028.	3.5	162
99	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
100	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
101	Maintenance of respiratory chain function in mouse hearts with severely impaired mtDNA transcription. Nucleic Acids Research, 2010, 38, 6577-6588.	14.5	35
102	Somatic Mitochondrial DNA Mutations in Mammalian Aging. Annual Review of Biochemistry, 2010, 79, 683-706.	11.1	456
103	LRPPRC is a mitochondrial matrix protein that is conserved in metazoans. Biochemical and Biophysical Research Communications, 2010, 398, 759-764.	2.1	49
104	Point Mutations Are Causing Progeroid Phenotypes in the mtDNA Mutator Mouse. Cell Metabolism, 2010, 11, 1.	16.2	12
105	Response: Point Mutations Are Causing Progeroid Phenotypes in the mtDNA Mutator Mouse. Cell Metabolism, 2010, 11, 93.	16.2	5
106	MTERF1 Gives mtDNA an Unusual Twist. Cell Metabolism, 2010, 12, 3-4.	16.2	3
107	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
108	Nurr1 Is Required for Maintenance of Maturing and Adult Midbrain Dopamine Neurons. Journal of Neuroscience, 2009, 29, 15923-15932.	3.6	320

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109	Increased mitochondrial Ca 2+ and decreased sarcoplasmic reticulum Ca 2+ in mitochondrial myopathy. Human Molecular Genetics, 2009, 18, 278-288.	2.9	64
110	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
111	MTERF2 is a nucleoid component in mammalian mitochondria. Biochimica Et Biophysica Acta - Bioenergetics, 2009, 1787, 296-302.	1.0	70
112	Introduction: Euromit VII â€“ summarizing 50â€ƒyears of research on mitochondrial disease. Journal of Internal Medicine, 2009, 265, 170-173.	6.0	0
113	Structure Casts Light on mtDNA Replication. Cell, 2009, 139, 231-233.	28.9	6
114	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
115	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
116	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
117	Introduction: Biology of ageing. Journal of Internal Medicine, 2008, 263, 114-116.	6.0	0
118	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
119	Purifying selection of mtDNA and its implications for understanding evolution and mitochondrial disease. Nature Reviews Genetics, 2008, 9, 657-662.	16.3	155
120	Mitochondrial dysfunction as a cause of ageing. Journal of Internal Medicine, 2008, 263, 167-178.	6.0	292
121	Complex I: A Complex Gateway to the Powerhouse. Cell Metabolism, 2008, 7, 278-279.	16.2	7
122	Age-associated mosaic respiratory chain deficiency causes trans-neuronal degeneration. Human Molecular Genetics, 2008, 17, 1418-1426.	2.9	41
123	Strong Purifying Selection in Transmission of Mammalian Mitochondrial DNA. PLoS Biology, 2008, 6, e10.	5.6	425
124	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
125	MTERF3 Is a Negative Regulator of Mammalian mtDNA Transcription. Cell, 2007, 130, 273-285.	28.9	209
126	Is Energy Deficiency Good in Moderation?. Cell, 2007, 131, 448-450.	28.9	13

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127	Somatic mtDNA mutations cause progressive hearing loss in the mouse. Experimental Cell Research, 2007, 313, 3924-3934.	2.6	48
128	DNA Replication and Transcription in Mammalian Mitochondria. Annual Review of Biochemistry, 2007, 76, 679-699.	11.1	567
129	Mitochondrial Dysfunction in Mammalian Ageing. Novartis Foundation Symposium, 2007, 287, 197-213.	1.1	63
130	Respiratory chain dysfunction in skeletal muscle does not cause insulin resistance. Biochemical and Biophysical Research Communications, 2006, 350, 202-207.	2.1	134
131	Proteolytic Processing of OPA1 Links Mitochondrial Dysfunction to Alterations in Mitochondrial Morphology. Journal of Biological Chemistry, 2006, 281, 37972-37979.	3.4	382
132	SOD2 overexpression: enhanced mitochondrial tolerance but absence of effect on UCP activity. EMBO Journal, 2005, 24, 4061-4070.	7.8	98
133	Secondary metabolic effects in complex I deficiency. Annals of Neurology, 2005, 58, 544-552.	5.3	51
134	A family of putative transcription termination factors shared amongst metazoans and plants. Current Genetics, 2005, 48, 265-269.	1.7	116
135	Abnormal Ca2+ release and catecholamine-induced arrhythmias in mitochondrial cardiomyopathy. Human Molecular Genetics, 2005, 14, 1069-1076.	2.9	22
136	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
137	Mitochondrial transcription factor A regulates mtDNA copy number in mammals. Human Molecular Genetics, 2004, 13, 935-944.	2.9	730
138	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
139	The mitochondrial RNA polymerase contributes critically to promoter specificity in mammalian cells. EMBO Journal, 2004, 23, 4606-4614.	7.8	151
140	Premature ageing in mice expressing defective mitochondrial DNA polymerase. Nature, 2004, 429, 417-423.	27.8	2,318
141	Understanding aging: revealing order out of chaos. Biochimica Et Biophysica Acta - Bioenergetics, 2004, 1658, 122-132.	1.0	80
142	The transcription machinery in mammalian mitochondria. Biochimica Et Biophysica Acta - Bioenergetics, 2004, 1659, 148-152.	1.0	94
143	Defective assembly of the respiratory chain. Acta Paediatrica, International Journal of Paediatrics, 2004, 93, 1268-1270.	1.5	0
144	Characterization of the mouse genes for mitochondrial transcription factors B1 and B2. Mammalian Genome, 2003, 14, 1-6.	2.2	34

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145	Breeding and Genotyping of Tfam Conditional Knockout Mice. , 2002, 197, 391-400.		13
146	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
147	Tissue-Specific Knockout Model for Study of Mitochondrial DNA Mutation Disorders. Methods in Enzymology, 2002, 353, 409-421.	1.0	9
148	Manipulation of mitochondrial DNA gene expression in the mouse. Biochimica Et Biophysica Acta - Bioenergetics, 2002, 1555, 106-110.	1.0	25
149	Mitochondrial diseases. Best Practice and Research in Clinical Obstetrics and Gynaecology, 2002, 16, 715-728.	2.8	17
150	Leber hereditary optic neuropathy: A nuclear solution of a mitochondrial problem. Annals of Neurology, 2002, 52, 529-530.	5.3	15
151	Mitochondrial transcription factors B1 and B2 activate transcription of human mtDNA. Nature Genetics, 2002, 31, 289-294.	21.4	535
152	Animal models for respiratory chain disease. Trends in Molecular Medicine, 2001, 7, 578-581.	6.7	44
153	Late-Onset Corticohippocampal Neurodepletion Attributable to Catastrophic Failure of Oxidative Phosphorylation in MILON Mice. Journal of Neuroscience, 2001, 21, 8082-8090.	3.6	151
154	Downregulation of Tfam and mtDNA copy number during mammalian spermatogenesis. Mammalian Genome, 2001, 12, 787-792.	2.2	66
155	Mitochondrial myopathies. Acta Physiologica Scandinavica, 2001, 171, 385-393.	2.2	54
156	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
157	Complex genetic counselling and prenatal analysis in a woman with external ophthalmoplegia and deleted mtDNA. , 2000, 20, 426-431.		18
158	Impaired insulin secretion and Î²-cell loss in tissue-specific knockout mice with mitochondrial diabetes. Nature Genetics, 2000, 26, 336-340.	21.4	417
159	Regulation of mitochondrial DNA copy number during spermatogenesis. Human Reproduction, 2000, 15, 86-91.	0.9	36
160	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
161	Mitochondrial medicine - recent advances. Journal of Internal Medicine, 1999, 246, 11-23.	6.0	70
162	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

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163	Revolution in mitochondrial medicine. FEBS Letters, 1999, 455, 199-202.	2.8	66
164	Mitochondrial transcription factor A is necessary for mtDNA maintance and embryogenesis in mice. Nature Genetics, 1998, 18, 231-236.	21.4	1,377
165	Down-Regulation of Mitochondrial Transcription Factor a During Spermatogenesis in Humans. Human Molecular Genetics, 1997, 6, 185-1991.	2.9	75
166	Fatal mitochondrial myopathy, lactic acidosis, and complex I deficiency associated with a heteroplasmic Aât'G mutation at position 3251 in the mitochondrial tRNALeu(UUR) gene. Human Genetics, 1996, 97, 269-273.	3.8	20
167	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
168	Mitochondrial DNA Deletions in Muscle Fibers in Inclusion Body Myositis. Journal of Neuropathology and Experimental Neurology, 1995, 54, 581-587.	1.7	103
169	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
170	Atypical presentation of multisystem disorders in two girls with mitochondrial DNA deletions. European Journal of Pediatrics, 1995, 154, 35-42.	2.7	30
171	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
172	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
173	MOLECULAR GENETIC ASPECTS OF HUMAN MITOCHONDRIAL DISORDERS. Annual Review of Genetics, 1995, 29, 151-178.	7.6	455
174	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
175	Mitochondrial DNA deletions in inclusion body myositis. Brain, 1993, 116, 325-336.	7.6	116
176	Mitochondrial ATP-Synthase Deficiency in a Child with 3-Methylglutaconic Aciduria. Pediatric Research, 1992, 32, 731-736.	2.3	65
177	Multiple short direct repeats associated with single mtDNA deletions. Biochimica Et Biophysica Acta - Molecular Basis of Disease, 1992, 1139, 311-314.	3.8	13
178	Mitochondrial DNA deletions and cytochrome c oxidase deficiency in muscle fibres. Journal of the Neurological Sciences, 1992, 110, 169-177.	0.6	61
179	Mitochondrial encephalomyopathies in childhood. I. Biochemical and morphologic investigations. Journal of Pediatrics, 1991, 119, 242-250.	1.8	128
180	Mitochondrial encephalomyopathies in childhood. II. Clinical manifestations and syndromes. Journal of Pediatrics, 1991, 119, 251-259.	1.8	85

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181	Leber's hereditary optic neuropathy and complex I deficiency in muscle. Annals of Neurology, 1991, 30, 701-708.	5.3	155
182	Progressive Increase of the Mutated Mitochondrial DNA Fraction in Kearns-Sayre Syndrome. Pediatric Research, 1990, 28, 131-136.	2.3	308
183	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
184	MitoTALEN reduces mutant mtDNA load and restores tRNAAla levels in a mouse model of heteroplasmic mtDNA mutation. , 0, .		1