Nils-Gran Larsson

List of Publications by Citations

183 papers	21,078 citations	73 h-index	144 g-index
188 ext. papers	23,926 ext. citations	13.3 avg, IF	6.87 L-index

#	Paper	IF	Citations
183	Premature ageing in mice expressing defective mitochondrial DNA polymerase. *Nature*, **2004**, 429, 417-23	50.4	1975
182	Mitochondrial transcription factor A is necessary for mtDNA maintenance and embryogenesis in mice. *Nature Genetics*, **1998**, 18, 231-6	36.3	1193
181	The role of mitochondria in aging. *Journal of Clinical Investigation*, **2013**, 123, 951-7	15.9	640
180	Mitochondrial transcription factor A regulates mtDNA copy number in mammals. *Human Molecular Genetics*, **2004**, 13, 935-44	5.6	599
179	Unraveling the biological roles of reactive oxygen species. *Cell Metabolism*, **2011**, 13, 361-366	24.6	542
178	DNA replication and transcription in mammalian mitochondria. *Annual Review of Biochemistry*, **2007** , 76, 679-99	29.1	488
177	Mitochondrial transcription factors B1 and B2 activate transcription of human mtDNA. *Nature Genetics*, **2002**, 31, 289-94	36.3	475
176	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, 1325-30	11.5	448
175	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, 17993-8	11.5	441
174	Molecular genetic aspects of human mitochondrial disorders. *Annual Review of Genetics*, **1995**, 29, 151-78	14.5	392
173	Somatic mitochondrial DNA mutations in mammalian aging. *Annual Review of Biochemistry*, **2010**, 79, 683-706	29.1	373
172	Impaired insulin secretion and beta-cell loss in tissue-specific knockout mice with mitochondrial diabetes. *Nature Genetics*, **2000**, 26, 336-40	36.3	371
171	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, 13534-9	11.5	360
170	Strong purifying selection in transmission of mammalian mitochondrial DNA. *PLoS Biology*, **2008**, 6, e10	9.7	352
169	Dilated cardiomyopathy and atrioventricular conduction blocks induced by heart-specific inactivation of mitochondrial DNA gene expression. *Nature Genetics*, **1999**, 21, 133-7	36.3	351
168	Maintenance and Expression of Mammalian Mitochondrial DNA. *Annual Review of Biochemistry*, **2016**, 85, 133-60	29.1	329
167	Proteolytic processing of OPA1 links mitochondrial dysfunction to alterations in mitochondrial morphology. *Journal of Biological Chemistry*, **2006**, 281, 37972-9	5.4	329

166	Mammalian Mitochondria and Aging: An Update. *Cell Metabolism*, **2017**, 25, 57-71	24.6	294
165	Progressive increase of the mutated mitochondrial DNA fraction in Kearns-Sayre syndrome. *Pediatric Research*, **1990**, 28, 131-6	3.2	277
164	Nurr1 is required for maintenance of maturing and adult midbrain dopamine neurons. *Journal of Neuroscience*, **2009**, 29, 15923-32	6.6	259
163	Mitochondrial dysfunction as a cause of ageing. *Journal of Internal Medicine*, **2008**, 263, 167-78	10.8	244
162	Methylation of 12S rRNA is necessary for in vivo stability of the small subunit of the mammalian mitochondrial ribosome. *Cell Metabolism*, **2009**, 9, 386-97	24.6	239
161	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, 12937-42	11.5	230
160	Increased mitochondrial mass in mitochondrial myopathy mice. *Proceedings of the National Academy of Sciences of the United States of America*, **2002**, 99, 15066-71	11.5	226
159	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, 4038-43	11.5	216
158	Mitochondrial DNA mutations in disease and aging. *Journal of Cell Biology*, **2011**, 193, 809-18	7.3	213
157	The Enigma of the Respiratory Chain Supercomplex. *Cell Metabolism*, **2017**, 25, 765-776	24.6	207
156	LRPPRC is necessary for polyadenylation and coordination of translation of mitochondrial mRNAs. *EMBO Journal*, **2012**, 31, 443-56	13	195
155	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, 11288-93	11.5	193
154	MTERF4 regulates translation by targeting the methyltransferase NSUN4 to the mammalian mitochondrial ribosome. *Cell Metabolism*, **2011**, 13, 527-39	24.6	190
153	Germline mitochondrial DNA mutations aggravate ageing and can impair brain development. *Nature*, **2013**, 501, 412-5	50.4	188
152	The role of mitochondrial DNA mutations and free radicals in disease and ageing. *Journal of Internal Medicine*, **2013**, 273, 529-43	10.8	185
151	Somatic progenitor cell vulnerability to mitochondrial DNA mutagenesis underlies progeroid phenotypes in Polg mutator mice. *Cell Metabolism*, **2012**, 15, 100-9	24.6	185
150	MTERF3 is a negative regulator of mammalian mtDNA transcription. *Cell*, **2007**, 130, 273-85	56.2	182
149	NSUN4 is a dual function mitochondrial protein required for both methylation of 12S rRNA and coordination of mitoribosomal assembly. *PLoS Genetics*, **2014**, 10, e1004110	6	179

148	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, 3136-41	11.5	178
147	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, 131-8	24.6	171
146	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, 20087-92	11.5	159
145	Adipose-specific deletion of TFAM increases mitochondrial oxidation and protects mice against obesity and insulin resistance. *Cell Metabolism*, **2012**, 16, 765-76	24.6	151
144	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, 3467-72	11.5	149
143	Making proteins in the powerhouse. *Cell Metabolism*, **2014**, 20, 226-40	24.6	139
142	Ultra-deep sequencing of mouse mitochondrial DNA: mutational patterns and their origins. *PLoS Genetics*, **2011**, 7, e1002028	6	136
141	Mitofusin 2 is required to maintain mitochondrial coenzyme Q levels. *Journal of Cell Biology*, **2015**, 208, 429-42	7.3	135
140	The mitochondrial RNA polymerase contributes critically to promoter specificity in mammalian cells. *EMBO Journal*, **2004**, 23, 4606-14	13	134
139	Mitofusin 2 is necessary for striatal axonal projections of midbrain dopamine neurons. *Human Molecular Genetics*, **2012**, 21, 4827-35	5.6	130
138	Late-onset corticohippocampal neurodepletion attributable to catastrophic failure of oxidative phosphorylation in MILON mice. *Journal of Neuroscience*, **2001**, 21, 8082-90	6.6	130
137	Purifying selection of mtDNA and its implications for understanding evolution and mitochondrial disease. *Nature Reviews Genetics*, **2008**, 9, 657-62	30.1	129
136	A single mouse gene encodes the mitochondrial transcription factor A and a testis-specific nuclear HMG-box protein. *Nature Genetics*, **1996**, 13, 296-302	36.3	129
135	Leber hereditary optic neuropathy and complex I deficiency in muscle. *Annals of Neurology*, **1991**, 30, 701-8	9.4	126
134	Respiratory chain dysfunction in skeletal muscle does not cause insulin resistance. *Biochemical and Biophysical Research Communications*, **2006**, 350, 202-7	3.4	122
133	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, 16510-5	11.5	118
132	MitoTALEN reduces mutant mtDNA load and restores tRNA levels in a mouse model of heteroplasmic mtDNA mutation. *Nature Medicine*, **2018**, 24, 1696-1700	50.5	117
131	Mitochondrial encephalomyopathies in childhood. I. Biochemical and morphologic investigations. *Journal of Pediatrics*, **1991**, 119, 242-50	3.6	116

130	TWINKLE is an essential mitochondrial helicase required for synthesis of nascent D-loop strands and complete mtDNA replication. *Human Molecular Genetics*, **2013**, 22, 1983-93	5.6	114
129	A family of putative transcription termination factors shared amongst metazoans and plants. *Current Genetics*, **2005**, 48, 265-9	2.9	108
128	Variation in germline mtDNA heteroplasmy is determined prenatally but modified during subsequent transmission. *Nature Genetics*, **2012**, 44, 1282-5	36.3	102
127	Mitochondrial DNA deletions in inclusion body myositis. *Brain*, **1993**, 116 (Pt 2), 325-36	11.2	98
126	Adipose tissue mitochondrial dysfunction triggers a lipodystrophic syndrome with insulin resistance, hepatosteatosis, and cardiovascular complications. *FASEB Journal*, **2014**, 28, 4408-19	0.9	95
125	Transcriptomic and proteomic landscape of mitochondrial dysfunction reveals secondary coenzyme Q deficiency in mammals. *ELife*, **2017**, 6,	8.9	93
124	Reactive oxygen species and fatigue-induced prolonged low-frequency force depression in skeletal muscle fibres of rats, mice and SOD2 overexpressing mice. *Journal of Physiology*, **2008**, 586, 175-84	3.9	92
123	SOD2 overexpression: enhanced mitochondrial tolerance but absence of effect on UCP activity. *EMBO Journal*, **2005**, 24, 4061-70	13	91
122	Impaired nigrostriatal function precedes behavioral deficits in a genetic mitochondrial model of Parkinson's disease. *FASEB Journal*, **2011**, 25, 1333-44	0.9	90
121	mtDNA makes a U-turn for the mitochondrial nucleoid. *Trends in Cell Biology*, **2013**, 23, 457-63	18.3	87
120	Structure of the human MTERF4-NSUN4 protein complex that regulates mitochondrial ribosome biogenesis. *Proceedings of the National Academy of Sciences of the United States of America*, **2012**, 109, 15253-8	11.5	87
119	The transcription machinery in mammalian mitochondria. *Biochimica Et Biophysica Acta - Bioenergetics*, **2004**, 1659, 148-52	4.6	85
118	MitoPark mice mirror the slow progression of key symptoms and L-DOPA response in Parkinson's disease. *Genes, Brain and Behavior*, **2010**, 9, 173-81	3.6	80
117	Hierarchical RNA Processing Is Required for Mitochondrial Ribosome Assembly. *Cell Reports*, **2016**, 16, 1874-90	10.6	80
116	MTERF1 binds mtDNA to prevent transcriptional interference at the light-strand promoter but is dispensable for rRNA gene transcription regulation. *Cell Metabolism*, **2013**, 17, 618-26	24.6	78
115	Loss of LRPPRC causes ATP synthase deficiency. *Human Molecular Genetics*, **2014**, 23, 2580-92	5.6	77
114	Mitochondrial DNA deletions in muscle fibers in inclusion body myositis. *Journal of Neuropathology and Experimental Neurology*, **1995**, 54, 581-7	3.1	77
113	Keeping mtDNA in shape between generations. *PLoS Genetics*, **2014**, 10, e1004670	6	76

112	Mitochondrial encephalomyopathies in childhood. II. Clinical manifestations and syndromes. *Journal of Pediatrics*, **1991**, 119, 251-9	3.6	75
111	AFG3L2 supports mitochondrial protein synthesis and Purkinje cell survival. *Journal of Clinical Investigation*, **2012**, 122, 4048-58	15.9	74
110	Changes of mitochondrial ultrastructure and function during ageing in mice and. *ELife*, **2017**, 6,	8.9	73
109	Understanding aging: revealing order out of chaos. *Biochimica Et Biophysica Acta - Bioenergetics*, **2004**, 1658, 122-32	4.6	72
108	A Phenotype-Driven Approach to Generate Mouse Models with Pathogenic mtDNA Mutations Causing Mitochondrial Disease. *Cell Reports*, **2016**, 16, 2980-2990	10.6	72
107	COX7A2L Is a Mitochondrial Complex III Binding Protein that Stabilizes the III2+IV Supercomplex without Affecting Respirasome Formation. *Cell Reports*, **2016**, 16, 2387-98	10.6	71
106	MTERF3 regulates mitochondrial ribosome biogenesis in invertebrates and mammals. *PLoS Genetics* , **2013**, 9, e1003178	6	70
105	The respiratory chain supercomplex organization is independent of COX7a2l isoforms. *Cell Metabolism*, **2014**, 20, 1069-75	24.6	69
104	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	24.6	64
103	MTERF2 is a nucleoid component in mammalian mitochondria. *Biochimica Et Biophysica Acta - Bioenergetics*, **2009**, 1787, 296-302	4.6	64
102	No recombination of mtDNA after heteroplasmy for 50 generations in the mouse maternal germline. *Nucleic Acids Research*, **2014**, 42, 1111-6	20.1	63
101	Down-regulation of mitochondrial transcription factor A during spermatogenesis in humans. *Human Molecular Genetics*, **1997**, 6, 185-91	5.6	63
100	Mitochondrial fusion is required for regulation of mitochondrial DNA replication. *PLoS Genetics*, **2019**, 15, e1008085	6	62
99	Topoisomerase 3β Is Required for Decatenation and Segregation of Human mtDNA. *Molecular Cell*, **2018**, 69, 9-23.e6	17.6	61
98	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, 1078-89	5.6	60
97	Mitochondrial ATP-synthase deficiency in a child with 3-methylglutaconic aciduria. *Pediatric Research*, **1992**, 32, 731-5	3.2	60
96	POLRMT regulates the switch between replication primer formation and gene expression of mammalian mtDNA. *Science Advances*, **2016**, 2, e1600963	14.3	58
95	Loss of the RNA-binding protein TACO1 causes late-onset mitochondrial dysfunction in mice. *Nature Communications*, **2016**, 7, 11884	17.4	57

94	Downregulation of Tfam and mtDNA copy number during mammalian spermatogenesis. *Mammalian Genome*, **2001**, 12, 787-92	3.2	56
93	Revolution in mitochondrial medicine. *FEBS Letters*, **1999**, 455, 199-202	3.8	56
92	Mitochondrial DNA deletions and cytochrome c oxidase deficiency in muscle fibres. *Journal of the Neurological Sciences*, **1992**, 110, 169-77	3.2	56
91	Mitochondrial dysfunction in mammalian ageing. *Novartis Foundation Symposium*, **2007**, 287, 197-208; discussion 208-13		56
90	A human mitochondrial poly(A) polymerase mutation reveals the complexities of post-transcriptional mitochondrial gene expression. *Human Molecular Genetics*, **2014**, 23, 6345-55	5.6	55
89	Increased mitochondrial Ca2+ and decreased sarcoplasmic reticulum Ca2+ in mitochondrial myopathy. *Human Molecular Genetics*, **2009**, 18, 278-88	5.6	55
88	Mitochondrial medicine--recent advances. *Journal of Internal Medicine*, **1999**, 246, 11-23	10.8	54
87	Mitochondrial DNA copy number in human disease: the more the better?. *FEBS Letters*, **2021**, 595, 976-1008	3.8	53
86	SLIRP Regulates the Rate of Mitochondrial Protein Synthesis and Protects LRPPRC from Degradation. *PLoS Genetics*, **2015**, 11, e1005423	6	52
85	Pathogenetic aspects of the A8344G mutation of mitochondrial DNA associated with MERRF syndrome and multiple symmetric lipomas. *Muscle and Nerve*, **1995**, 3, S102-6	3.4	52
84	LRPPRC-mediated folding of the mitochondrial transcriptome. *Nature Communications*, **2017**, 8, 1532	17.4	50
83	In vivo mutagenesis reveals that OriL is essential for mitochondrial DNA replication. *EMBO Reports*, **2012**, 13, 1130-7	6.5	49
82	Increased Total mtDNA Copy Number Cures Male Infertility Despite Unaltered mtDNA Mutation Load. *Cell Metabolism*, **2017**, 26, 429-436.e4	24.6	46
81	The bicoid stability factor controls polyadenylation and expression of specific mitochondrial mRNAs in Drosophila melanogaster. *PLoS Genetics*, **2011**, 7, e1002324	6	46
80	Mitochondrial myopathies. *Acta Physiologica Scandinavica*, **2001**, 171, 385-93		45
79	LRPPRC is a mitochondrial matrix protein that is conserved in metazoans. *Biochemical and Biophysical Research Communications*, **2010**, 398, 759-64	3.4	44
78	Somatic mtDNA mutations cause progressive hearing loss in the mouse. *Experimental Cell Research*, **2007**, 313, 3924-34	4.2	44
77	Secondary metabolic effects in complex I deficiency. *Annals of Neurology*, **2005**, 58, 544-52	9.4	43

76	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	20.1	41
75	The mitochondrial electron transport chain is dispensable for proliferation and differentiation of epidermal progenitor cells. *Stem Cells*, **2011**, 29, 1459-68	5.8	41
74	Age-associated mosaic respiratory chain deficiency causes trans-neuronal degeneration. *Human Molecular Genetics*, **2008**, 17, 1418-26	5.6	40
73	Animal models for respiratory chain disease. *Trends in Molecular Medicine*, **2001**, 7, 578-81	11.5	40
72	Bioenergetic roles of mitochondrial fusion. *Biochimica Et Biophysica Acta - Bioenergetics*, **2016**, 1857, 1277-1283	4.6	39
71	Mice lacking the mitochondrial exonuclease MGME1 accumulate mtDNA deletions without developing progeria. *Nature Communications*, **2018**, 9, 1202	17.4	38
70	MtDNA mutations are a common cause of severe disease phenotypes in children with Leigh syndrome. *Biochimica Et Biophysica Acta - Bioenergetics*, **2009**, 1787, 484-90	4.6	38
69	Loss of TFB1M results in mitochondrial dysfunction that leads to impaired insulin secretion and diabetes. *Human Molecular Genetics*, **2014**, 23, 5733-49	5.6	37
68	Automatic sequencing of mitochondrial tRNA genes in patients with mitochondrial encephalomyopathy. *Biochimica Et Biophysica Acta - Molecular Basis of Disease*, **1994**, 1226, 49-55	6.9	37
67	The amino terminal extension of mammalian mitochondrial RNA polymerase ensures promoter specific transcription initiation. *Nucleic Acids Research*, **2014**, 42, 3638-47	20.1	36
66	Two novel mutations in thymidine kinase-2 cause early onset fatal encephalomyopathy and severe mtDNA depletion. *Neuromuscular Disorders*, **2010**, 20, 198-203	2.9	36
65	Complementation between polymerase- and exonuclease-deficient mitochondrial DNA polymerase mutants in genomically engineered flies. *Nature Communications*, **2015**, 6, 8808	17.4	35
64	Modulation of mtDNA copy number ameliorates the pathological consequences of a heteroplasmic mtDNA mutation in the mouse. *Science Advances*, **2019**, 5, eaav9824	14.3	33
63	Small-molecule inhibitors of human mitochondrial DNA transcription. *Nature*, **2020**, 588, 712-716	50.4	32
62	Characterization of the mouse genes for mitochondrial transcription factors B1 and B2. *Mammalian Genome*, **2003**, 14, 1-6	3.2	32
61	POLRMT does not transcribe nuclear genes. *Nature*, **2014**, 514, E7-11	50.4	31
60	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	6.3	31
59	SnapShot: Mitochondrial Nucleoid. *Cell*, **2018**, 172, 388-388.e1	56.2	30

58	Sensory ataxic neuropathy in golden retriever dogs is caused by a deletion in the mitochondrial tRNATyr gene. *PLoS Genetics*, **2009**, 5, e1000499	6	30
57	PTCD1 Is Required for 16S rRNA Maturation Complex Stability and Mitochondrial Ribosome Assembly. *Cell Reports*, **2018**, 23, 127-142	10.6	29
56	Maintenance of respiratory chain function in mouse hearts with severely impaired mtDNA transcription. *Nucleic Acids Research*, **2010**, 38, 6577-88	20.1	29
55	Defects in β-cell Ca2+ dynamics in age-induced diabetes. *Diabetes*, **2014**, 63, 4100-14	0.9	26
54	Atypical presentation of multisystem disorders in two girls with mitochondrial DNA deletions. *European Journal of Pediatrics*, **1995**, 154, 35-42	4.1	26
53	SLIRP stabilizes LRPPRC via an RRM-PPR protein interface. *Nucleic Acids Research*, **2016**, 44, 6868-82	20.1	24
52	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	5.6	23
51	Sequence-specific stalling of DNA polymerase γ and the effects of mutations causing progressive ophthalmoplegia. *Human Molecular Genetics*, **2011**, 20, 1212-23	5.6	23
50	Regulation of mitochondrial DNA copy number during spermatogenesis. *Human Reproduction*, **2000**, 15 Suppl 2, 86-91	5.7	23
49	TEFM regulates both transcription elongation and RNA processing in mitochondria. *EMBO Reports*, **2019**, 20,	6.5	22
48	The leucine-rich pentatricopeptide repeat-containing protein (LRPPRC) does not activate transcription in mammalian mitochondria. *Journal of Biological Chemistry*, **2013**, 288, 15510-9	5.4	22
47	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	11.5	22
46	Abnormal Ca(2+) release and catecholamine-induced arrhythmias in mitochondrial cardiomyopathy. *Human Molecular Genetics*, **2005**, 14, 1069-76	5.6	20
45	Manipulation of mitochondrial DNA gene expression in the mouse. *Biochimica Et Biophysica Acta - Bioenergetics*, **2002**, 1555, 106-10	4.6	20
44	Drosophila melanogaster LRPPRC2 is involved in coordination of mitochondrial translation. *Nucleic Acids Research*, **2014**, 42, 13920-38	20.1	18
43	Tracing the trail of protons through complex I of the mitochondrial respiratory chain. *PLoS Biology*, **2011**, 9, e1001129	9.7	18
42	Cellular pyrimidine imbalance triggers mitochondrial DNA-dependent innate immunity. *Nature Metabolism*, **2021**, 3, 636-650	14.6	17
41	Fatal mitochondrial myopathy, lactic acidosis, and complex I deficiency associated with a heteroplasmic A --> G mutation at position 3251 in the mitochondrial tRNALeu(UUR) gne. *Human Genetics*, **1996**, 97, 269-73	6.3	16

40	Dinucleotide Degradation by REXO2 Maintains Promoter Specificity in Mammalian Mitochondria. *Molecular Cell*, **2019**, 76, 784-796.e6	17.6	14
39	Mitochondrial diseases. *Best Practice and Research in Clinical Obstetrics and Gynaecology*, **2002**, 16, 715-28	4.6	14
38	Complex genetic counselling and prenatal analysis in a woman with external ophthalmoplegia and deleted mtDNA **2000**, 20, 426-431		14
37	Mic10 Oligomerization Pinches off Mitochondrial Cristae. *Cell Metabolism*, **2015**, 21, 660-1	24.6	13
36	MitoRibo-Tag Mice Provide a Tool for In Vivo Studies of Mitoribosome Composition. *Cell Reports*, **2019**, 29, 1728-1738.e9	10.6	12
35	Neuronal metabolic rewiring promotes resilience to neurodegeneration caused by mitochondrial dysfunction. *Science Advances*, **2020**, 6, eaba8271	14.3	12
34	Point mutations are causing progeroid phenotypes in the mtDNA mutator mouse. *Cell Metabolism*, **2010**, 11, 1	24.6	11
33	Is energy deficiency good in moderation?. *Cell*, **2007**, 131, 448-50	56.2	11
32	Breeding and genotyping of Tfam conditional knockout mice. *Methods in Molecular Biology*, **2002**, 197, 391-400	1.4	11
31	Accurate mapping of mitochondrial DNA deletions and duplications using deep sequencing. *PLoS Genetics*, **2020**, 16, e1009242	6	11
30	FBXL4 deficiency increases mitochondrial removal by autophagy. *EMBO Molecular Medicine*, **2020**, 12, e11659	12	11
29	Leber hereditary optic neuropathy: a nuclear solution of a mitochondrial problem. *Annals of Neurology*, **2002**, 52, 529-30	9.4	10
28	Multiple short direct repeats associated with single mtDNA deletions. *Biochimica Et Biophysica Acta - Molecular Basis of Disease*, **1992**, 1139, 311-4	6.9	10
27	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, 389-95	7.5	10
26	Enhanced cardiomyocyte Ca(2+) cycling precedes terminal AV-block in mitochondrial cardiomyopathy Mterf3 KO mice. *Antioxidants and Redox Signaling*, **2011**, 15, 2455-64	8.4	9
25	The mitochondrial single-stranded DNA binding protein is essential for initiation of mtDNA replication. *Science Advances*, **2021**, 7,	14.3	9
24	Tissue-specific knockout model for study of mitochondrial DNA mutation disorders. *Methods in Enzymology*, **2002**, 353, 409-21	1.7	8
23	Overexpression of the mitochondrial methyltransferase TFB1M in the mouse does not impact mitoribosomal methylation status or hearing. *Human Molecular Genetics*, **2015**, 24, 7286-94	5.6	7

22	The UbL protein UBTD1 stably interacts with the UBE2D family of E2 ubiquitin conjugating enzymes. *Biochemical and Biophysical Research Communications*, **2014**, 443, 7-12	3.4	6
21	High levels of TFAM repress mammalian mitochondrial DNA transcription in vivo. *Life Science Alliance*, **2021**, 4,	5.8	6
20	Structure casts light on mtDNA replication. *Cell*, **2009**, 139, 231-3	56.2	5
19	Complex I: a complex gateway to the powerhouse. *Cell Metabolism*, **2008**, 7, 278-9	24.6	5
18	An Adaptable High-Throughput Technology Enabling the Identification of Specific Transcription Modulators. *SLAS Discovery*, **2017**, 22, 378-386	3.4	4
17	Response: Point Mutations Are Causing Progeroid Phenotypes in the mtDNA Mutator Mouse. *Cell Metabolism*, **2010**, 11, 93	24.6	4
16	Proofreading deficiency in mitochondrial DNA polymerase does not affect total dNTP pools in mouse embryos. *Nature Metabolism*, **2020**, 2, 673-675	14.6	4
15	MTERF1 gives mtDNA an unusual twist. *Cell Metabolism*, **2010**, 12, 3-4	24.6	3
14	Complete Deletion of a POLG1 Allele in a Patient with Alpers Syndrome. *JIMD Reports*, **2012**, 4, 67-73	1.9	3
13	Use of primary cultures and continuous cell lines to study effects on astrocytic regulatory functions. *Clinical and Experimental Pharmacology and Physiology*, **1995**, 22, 284-7	3	3
12	Mitochondria in human disease. *Journal of Internal Medicine*, **2020**, 287, 589-591	10.8	1
11	MitoTALEN reduces mutant mtDNA load and restores tRNAAla levels in a mouse model of heteroplasmic mtDNA mutation		1
10	Metabolic resistance to the inhibition of mitochondrial transcription revealed by CRISPR-Cas9 screen. *EMBO Reports*, **2021**, e53054	6.5	0
9	Mitochondrial dysfunction in adult midbrain dopamine neurons triggers an early immune response. *PLoS Genetics*, **2021**, 17, e1009822	6	0
8	Mice lacking the mitochondrial exonuclease MGME1 develop inflammatory kidney disease with glomerular dysfunction.. *PLoS Genetics*, **2022**, 18, e1010190	6	0
7	Introduction: Euromit VII--summarizing 50 years of research on mitochondrial disease. *Journal of Internal Medicine*, **2009**, 265, 170-3	10.8	
6	Defective assembly of the respiratory chain. *Acta Paediatrica, International Journal of Paediatrics*, **2007**, 93, 1268-1270	3.1	
5	Introduction: Biology of ageing. *Journal of Internal Medicine*, **2008**, 263, 114-116	10.8	

4 Accurate mapping of mitochondrial DNA deletions and duplications using deep sequencing **2020**, 16, e1009242

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