MichaÅ, MiÅ„czuk

List of Publications by Year
in descending order

94 papers	6,580 citations	41344 49 h-index	71685 76 g-index
108 all docs	108 docs citations	108 times ranked	7413 citing authors

#	Article	IF	Citations
1	Mitochondrially targeted <scp>ZFN</scp>s for selective degradation of pathogenic mitochondrial genomes bearing largeâ€scale deletions or point mutations. EMBO Molecular Medicine, 2014, 6, 458-466.	6.9	237
2	Mitochondrial Genome Engineering: The Revolution May Not Be CRISPR-Ized. Trends in Genetics, 2018, 34, 101-110.	6.7	230
3	Genome editing in mitochondria corrects a pathogenic mtDNA mutation in vivo. Nature Medicine, 2018, 24, 1691-1695.	30.7	215
4	In D-loop: 40years of mitochondrial 7S DNA. Experimental Gerontology, 2014, 56, 175-181.	2.8	207
5	Loss-of-function mutations in MGME1 impair mtDNA replication and cause multisystemic mitochondrial disease. Nature Genetics, 2013, 45, 214-219.	21.4	198
6	Development of a single-chain, quasi-dimeric zinc-finger nuclease for the selective degradation of mutated human mitochondrial DNA. Nucleic Acids Research, 2008, 36, 3926-3938.	14.5	195
7	Mitochondrial transcription and translation: overview. Essays in Biochemistry, 2018, 62, 309-320.	4.7	192
8	Deficient methylation and formylation of mt-tRNAMet wobble cytosine in a patient carrying mutations in NSUN3. Nature Communications, 2016, 7, 12039.	12.8	178
9	NADH Shuttling Couples Cytosolic Reductive Carboxylation of Glutamine with Glycolysis in Cells with Mitochondrial Dysfunction. Molecular Cell, 2018, 69, 581-593.e7.	9.7	171
10	Regulation of Mammalian Mitochondrial Gene Expression: Recent Advances. Trends in Biochemical Sciences, 2017, 42, 625-639.	7.5	151
11	Linear mitochondrial DNA is rapidly degraded by components of the replication machinery. Nature Communications, 2018, 9, 1727.	12.8	151
12	Sequence-specific modification of mitochondrial DNA using a chimeric zinc finger methylase. Proceedings of the National Academy of Sciences of the United States of America, 2006, 103, 19689-19694.	7.1	147
13	TEFM (c17orf42) is necessary for transcription of human mtDNA. Nucleic Acids Research, 2011, 39, 4284-4299.	14.5	142
14	The Yeast Mitochondrial Degradosome. Journal of Biological Chemistry, 2003, 278, 1603-1611.	3.4	135
15	Designer zinc-finger proteins and their applications. Gene, 2006, 366, 27-38.	2.2	131
16	ELAC2 Mutations Cause a Mitochondrial RNA Processing Defect Associated with Hypertrophic Cardiomyopathy. American Journal of Human Genetics, 2013, 93, 211-223.	6.2	127
17	Chimeric DNA methyltransferases target DNA methylation to specific DNA sequences and repress expression of target genes. Nucleic Acids Research, 2007, 35, 100-112.	14.5	126
18	Mutations in GTPBP3 Cause a Mitochondrial Translation Defect Associated with Hypertrophic Cardiomyopathy, Lactic Acidosis, and Encephalopathy. American Journal of Human Genetics, 2014, 95, 708-720.	6.2	123

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19	The post-transcriptional life of mammalian mitochondrial RNA. Biochemical Journal, 2012, 444, 357-373.	3.7	114
20	MRM2 and MRM3 are involved in biogenesis of the large subunit of the mitochondrial ribosome. Molecular Biology of the Cell, 2014, 25, 2542-2555.	2.1	99
21	Near-complete elimination of mutant mtDNA by iterative or dynamic dose-controlled treatment with mtZFNs. Nucleic Acids Research, 2016, 44, 7804-7816.	14.5	97
22	PDE12 removes mitochondrial RNA poly(A) tails and controls translation in human mitochondria. Nucleic Acids Research, 2011, 39, 7750-7763.	14.5	91
23	Variants in PUS7 Cause Intellectual Disability with Speech Delay, Microcephaly, Short Stature, and Aggressive Behavior. American Journal of Human Genetics, 2018, 103, 1045-1052.	6.2	89
24	VARS2andTARS2Mutations in Patients with Mitochondrial Encephalomyopathies. Human Mutation, 2014, 35, 983-989.	2.5	86
25	NSUN2 introduces 5-methylcytosines in mammalian mitochondrial tRNAs. Nucleic Acids Research, 2019, 47, 8720-8733.	14.5	84
26	An Upstream Open Reading Frame and the Context of the Two AUG Codons Affect the Abundance of Mitochondrial and Nuclear RNase H1. Molecular and Cellular Biology, 2010, 30, 5123-5134.	2.3	83
27	TRMT5 Mutations Cause a Defect in Post-transcriptional Modification of Mitochondrial tRNA Associated with Multiple Respiratory-Chain Deficiencies. American Journal of Human Genetics, 2015, 97, 319-328.	6.2	83
28	Localisation of the human hSuv3p helicase in the mitochondrial matrix and its preferential unwinding of dsDNA. Nucleic Acids Research, 2002, 30, 5074-5086.	14.5	81
29	The Pseudouridine Synthase RPUSD4 Is an Essential Component of Mitochondrial RNA Granules. Journal of Biological Chemistry, 2017, 292, 4519-4532.	3.4	79
30	Human Polynucleotide Phosphorylase, hPNPase, is Localized in Mitochondria. Journal of Molecular Biology, 2003, 329, 853-857.	4.2	78
31	Myosin VI-Dependent Actin Cages Encapsulate Parkin-Positive Damaged Mitochondria. Developmental Cell, 2018, 44, 484-499.e6.	7.0	77
32	Inhibition of herpes simplex virus 1 gene expression by designer zinc-finger transcription factors. Proceedings of the National Academy of Sciences of the United States of America, 2003, 100, 1621-1626.	7.1	76
33	Elongational stalling activates mitoribosome-associated quality control. Science, 2020, 370, 1105-1110.	12.6	74
34	Maturation of selected human mitochondrial tRNAs requires deadenylation. ELife, 2017, 6, .	6.0	72
35	METTL15 introduces N4-methylcytidine into human mitochondrial 12S rRNA and is required for mitoribosome biogenesis. Nucleic Acids Research, 2019, 47, 10267-10281.	14.5	70
36	Mitochondrial transcript maturation and its disorders. Journal of Inherited Metabolic Disease, 2015, 38, 655-680.	3.6	69

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37	Nuclear-encoded factors involved in post-transcriptional processing and modification of mitochondrial tRNAs in human disease. Frontiers in Genetics, 2015, 6, 79.	2.3	69
38	Potent Inhibition of NTPase/Helicase of the West Nile Virus by Ring-Expanded (â€œFatâ€) Nucleoside Analogues. Journal of Medicinal Chemistry, 2003, 46, 4776-4789.	6.4	68
39	C7orf30 is necessary for biogenesis of the large subunit of the mitochondrial ribosome. Nucleic Acids Research, 2012, 40, 4097-4109.	14.5	64
40	Linear mtDNA fragments and unusual mtDNA rearrangements associated with pathological deficiency of MGME1 exonuclease. Human Molecular Genetics, 2014, 23, 6147-6162.	2.9	64
41	TRNT1 deficiency: clinical, biochemical and molecular genetic features. Orphanet Journal of Rare Diseases, 2016, 11, 90.	2.7	64
42	Human mitochondrial ribosomes can switch their structural RNA composition. Proceedings of the National Academy of Sciences of the United States of America, 2016, 113, 12198-12201.	7.1	64
43	The yeast nuclear gene DSS1, which codes for a putative RNase II, is necessary for the function of the mitochondrial degradosome in processing and turnover of RNA. Molecular Genetics and Genomics, 1998, 260, 108-114.	2.4	63
44	Defective mitochondrial rRNA methyltransferase MRM2 causes MELAS-like clinical syndrome. Human Molecular Genetics, 2017, 26, 4257-4266.	2.9	63
45	Mutations in the mitochondrial cysteinyl-tRNA synthase gene,CARS2,lead to a severe epileptic encephalopathy and complex movement disorder. Journal of Medical Genetics, 2015, 52, 532-540.	3.2	62
46	Mechanisms of Mitochondrial DNA Deletion Formation. Trends in Genetics, 2019, 35, 235-244.	6.7	62
47	The potential of mitochondrial genome engineering. Nature Reviews Genetics, 2022, 23, 199-214.	16.3	59
48	Biallelic C1QBP Mutations Cause Severe Neonatal-, Childhood-, or Later-Onset Cardiomyopathy Associated with Combined Respiratory-Chain Deficiencies. American Journal of Human Genetics, 2017, 101, 525-538.	6.2	58
49	Alternative translation initiation augments the human mitochondrial proteome. Nucleic Acids Research, 2013, 41, 2354-2369.	14.5	56
50	MPV17L2 is required for ribosome assembly in mitochondria. Nucleic Acids Research, 2014, 42, 8500-8515.	14.5	56
51	Human Cytomegalovirus Infection Upregulates the Mitochondrial Transcription and Translation Machineries. MBio, 2016, 7, e00029.	4.1	55
52	Therapeutic Manipulation of mtDNA Heteroplasmy: A Shifting Perspective. Trends in Molecular Medicine, 2020, 26, 698-709.	6.7	52
53	Two Siblings with Homozygous Pathogenic Splice-Site Variant in Mitochondrial Asparaginyl-tRNA Synthetase (NARS2). Human Mutation, 2015, 36, 222-231.	2.5	51
54	Construction and testing of engineered zinc-finger proteins for sequence-specific modification of mtDNA. Nature Protocols, 2010, 5, 342-356.	12.0	50

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55	Amino Acid Starvation Has Opposite Effects on Mitochondrial and Cytosolic Protein Synthesis. PLoS ONE, 2014, 9, e93597.	2.5	48
56	TRMT2B is responsible for both tRNA and rRNA m⁵U-methylation in human mitochondria. RNA Biology, 2020, 17, 451-462.	3.1	46
57	In vivo mitochondrial base editing via adeno-associated viral delivery to mouse post-mitotic tissue. Nature Communications, 2022, 13, 750.	12.8	45
58	Pathogenic variants in glutamyl-tRNAGln amidotransferase subunits cause a lethal mitochondrial cardiomyopathy disorder. Nature Communications, 2018, 9, 4065.	12.8	44
59	Balancing of mitochondrial translation through METTL8-mediated m3C modification of mitochondrial tRNAs. Molecular Cell, 2021, 81, 4810-4825.e12.	9.7	44
60	Disruption of the TCA cycle reveals an ATF4-dependent integration of redox and amino acid metabolism. ELife, 2021, 10, .	6.0	44
61	The mammalian mitochondrial epitranscriptome. Biochimica Et Biophysica Acta - Gene Regulatory Mechanisms, 2019, 1862, 429-446.	1.9	40
62	New insights into the phenotype of FARS2 deficiency. Molecular Genetics and Metabolism, 2017, 122, 172-181.	1.1	38
63	Human ATP-dependent RNA/DNA helicase hSuv3p interacts with the cofactor of survivin HBXIP. FEBS Journal, 2005, 272, 5008-5019.	4.7	37
64	Mitochondria: Mitochondrial RNA metabolism and human disease. International Journal of Biochemistry and Cell Biology, 2013, 45, 845-849.	2.8	34
65	Engineered mtZFNs for Manipulation of Human Mitochondrial DNA Heteroplasmy. Methods in Molecular Biology, 2016, 1351, 145-162.	0.9	33
66	Mutations inELAC2associated with hypertrophic cardiomyopathy impair mitochondrial tRNA 3â€²â€end processing. Human Mutation, 2019, 40, 1731-1748.	2.5	31
67	Macropinocytic entry of isolated mitochondria in epidermal growth factor-activated human osteosarcoma cells. Scientific Reports, 2017, 7, 12886.	3.3	30
68	The structure of human EXD2 reveals a chimeric 3â€² to 5â€² exonuclease domain that discriminates substrates via metal coordination. Nucleic Acids Research, 2019, 47, 7078-7093.	14.5	29
69	A homozygous MRPL24 mutation causes a complex movement disorder and affects the mitoribosome assembly. Neurobiology of Disease, 2020, 141, 104880.	4.4	29
70	EXD2 Protects Stressed Replication Forks and Is Required for Cell Viability in the Absence of BRCA1/2. Molecular Cell, 2019, 75, 605-619.e6.	9.7	26
71	Metabolic shift underlies recovery in reversible infantile respiratory chain deficiency. EMBO Journal, 2020, 39, e105364.	7.8	26
72	DealingÂwithÂanÂUnconventionalÂGeneticÂCodeÂinÂ Mitochondria:ÂTheÂBiogenesisÂandÂPathogenicÂ DefectsÂofÂtheÂ5â€²FormylcytosineÂModificationÂinÂ MitochondrialÂtRNAMet. Biomolecules, 2017, 7, 24.	4.0	24

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73	Polyadenylation in Bacteria and Organelles. Methods in Molecular Biology, 2014, 1125, 211-227.	0.9	23
74	Mitochondrially-targeted APOBEC1 is a potent mtDNA mutator affecting mitochondrial function and organismal fitness in Drosophila. Nature Communications, 2019, 10, 3280.	12.8	23
75	Energetic costs of cellular and therapeutic control of stochastic mitochondrial DNA populations. PLoS Computational Biology, 2019, 15, e1007023.	3.2	20
76	Heterozygous SSBP1 start loss mutation co-segregates with hearing loss and the m.1555A>G mtDNA variant in a large multigenerational family. Brain, 2018, 141, 55-62.	7.6	19
77	Overexpressed yeast mitochondrial putative RNA helicase Mss116 partially restores proper mtRNA metabolism in strains lacking the Suv3 mtRNA helicase. Yeast, 2002, 19, 1285-1293.	1.7	18
78	Human mitochondrial ribosomes can switch structural tRNAs â€“ but when and why?. RNA Biology, 2017, 14, 1668-1671.	3.1	18
79	Quantitative density gradient analysis by mass spectrometry (qDGMS) and complexome profiling analysis (ComPrAn) R package for the study of macromolecular complexes. Biochimica Et Biophysica Acta - Bioenergetics, 2021, 1862, 148399.	1.0	16
80	The FASTK family proteins fine-tune mitochondrial RNA processing. PLoS Genetics, 2021, 17, e1009873.	3.5	16
81	Manipulation of mitochondrial genes and mtDNA heteroplasmy. Methods in Cell Biology, 2020, 155, 441-487.	1.1	15
82	DNA polymerase gamma mutations that impair holoenzyme stability cause catalytic subunit depletion. Nucleic Acids Research, 2021, 49, 5230-5248.	14.5	15
83	Duplexing complexome profiling with SILAC to study human respiratory chain assembly defects. Biochimica Et Biophysica Acta - Bioenergetics, 2021, 1862, 148395.	1.0	15
84	A late-stage assembly checkpoint of the human mitochondrial ribosome large subunit. Nature Communications, 2022, 13, 929.	12.8	13
85	The 5â€² region of the human hSUV3 gene encoding mitochondrial DNA and RNA helicase: Promoter characterization and alternative pre-mRNA splicing. Biochimica Et Biophysica Acta Gene Regulatory Mechanisms, 2005, 1729, 81-87.	2.4	11
86	Engineered Zinc Finger Proteins for Manipulation of the Human Mitochondrial Genome. Methods in Molecular Biology, 2010, 649, 257-270.	0.9	10
87	Cardiac mitochondrial function depends on BUD23 mediated ribosome programming. ELife, 2020, 9, .	6.0	10
88	Enhanced Manipulation of Human Mitochondrial DNA Heteroplasmy In Vitro Using Tunable mtZFN Technology. Methods in Molecular Biology, 2018, 1867, 43-56.	0.9	8
89	YbeY is required for ribosome small subunit assembly and tRNA processing in human mitochondria. Nucleic Acids Research, 2021, 49, 5798-5812.	14.5	8
90	Detection of 5-formylcytosine in Mitochondrial Transcriptome. Methods in Molecular Biology, 2021, 2192, 59-68.	0.9	7

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91	Delivery of mtZFNs into Early Mouse Embryos. Methods in Molecular Biology, 2018, 1867, 215-228.	0.9	6
92	RNA Degradation in Yeast and Human Mitochondria. Toxicology Mechanisms and Methods, 2004, 14, 53-57.	2.7	1
93	Polyadenylation of mt mRNA: Identification of novel deadenylase of human mitochondria. Biochimica Et Biophysica Acta - Bioenergetics, 2010, 1797, 105.	1.0	0
94	Mitochondrially targeted zinc finger nucleases. , 2020, , 499-514.		0