# Jan W Taanman

## List of Publications by Citations



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|---|---|---|---|
| 117 papers | 7,518 citations | 43 h-index | 86 g-index |
| 126 ext. papers | 8,205 ext. citations | 6.1 avg, IF | 5.72 L-index |

| # | Paper | IF | Citations |
|---|---|---|---|
| 117 | The mitochondrial genome: structure, transcription, translation and replication. *Biochimica Et Biophysica Acta - Bioenergetics*, **1999**, 1410, 103-23 | 4.6 | 940 |
| 116 | Mitofusin 1 and mitofusin 2 are ubiquitinated in a PINK1/parkin-dependent manner upon induction of mitophagy. *Human Molecular Genetics*, **2010**, 19, 4861-70 | 5.6 | 680 |
| 115 | Biochemical abnormalities and excitotoxicity in Huntington's disease brain. *Annals of Neurology*, **1999**, 45, 25-32 | 9.4 | 393 |
| 114 | Parkinson's disease induced pluripotent stem cells with triplication of the □synuclein locus. *Nature Communications*, **2011**, 2, 440 | 17.4 | 328 |
| 113 | Mitochondrial DNA transmission of the mitochondrial defect in Parkinson's disease. *Annals of Neurology*, **1998**, 44, 177-86 | 9.4 | 278 |
| 112 | A mutant mitochondrial respiratory chain assembly protein causes complex III deficiency in patients with tubulopathy, encephalopathy and liver failure. *Nature Genetics*, **2001**, 29, 57-60 | 36.3 | 262 |
| 111 | Cytochrome c oxidase subunit I microdeletion in a patient with motor neuron disease. *Annals of Neurology*, **1998**, 43, 110-6 | 9.4 | 226 |
| 110 | A mutation in the human heme A:farnesyltransferase gene (COX10 ) causes cytochrome c oxidase deficiency. *Human Molecular Genetics*, **2000**, 9, 1245-9 | 5.6 | 223 |
| 109 | Assembly of cytochrome-c oxidase in cultured human cells. *FEBS Journal*, **1998**, 254, 389-94 | | 193 |
| 108 | Expression of mutant alpha-synuclein causes increased susceptibility to dopamine toxicity. *Human Molecular Genetics*, **2000**, 9, 2683-9 | 5.6 | 166 |
| 107 | Silencing of PINK1 expression affects mitochondrial DNA and oxidative phosphorylation in dopaminergic cells. *PLoS ONE*, **2009**, 4, e4756 | 3.7 | 147 |
| 106 | Dominant inheritance of premature ovarian failure associated with mutant mitochondrial DNA polymerase gamma. *Human Reproduction*, **2006**, 21, 2467-73 | 5.7 | 132 |
| 105 | Status epilepticus in children with Alpers' disease caused by POLG1 mutations: EEG and MRI features. *Epilepsia*, **2009**, 50, 1596-607 | 6.4 | 123 |
| 104 | A missense mutation of cytochrome oxidase subunit II causes defective assembly and myopathy. *American Journal of Human Genetics*, **1999**, 65, 1030-9 | 11 | 120 |
| 103 | Molecular mechanisms in mitochondrial DNA depletion syndrome. *Human Molecular Genetics*, **1997**, 6, 935-42 | 5.6 | 112 |
| 102 | Mammalian cytochrome-c oxidase: characterization of enzyme and immunological detection of subunits in tissue extracts and whole cells. *Methods in Enzymology*, **1995**, 260, 117-32 | 1.7 | 109 |
| 101 | Cytochrome c oxidase subassemblies in fibroblast cultures from patients carrying mutations in COX10, SCO1, or SURF1. *Journal of Biological Chemistry*, **2004**, 279, 7462-9 | 5.4 | 102 |

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|---|---|---|---|
| 100 | Liver failure associated with mitochondrial DNA depletion. *Journal of Hepatology*, **1998**, 28, 556-63 | 13.4 | 95 |
| 99 | A novel frameshift mutation of the mtDNA COIII gene leads to impaired assembly of cytochrome c oxidase in a patient affected by Leigh-like syndrome. *Human Molecular Genetics*, **2000**, 9, 2733-42 | 5.6 | 93 |
| 98 | Differential effects of PINK1 nonsense and missense mutations on mitochondrial function and morphology. *Experimental Neurology*, **2009**, 219, 266-73 | 5.7 | 90 |
| 97 | Expression of mtDNA and nDNA encoded respiratory chain proteins in chemically and genetically-derived Rho0 human fibroblasts: a comparison of subunit proteins in normal fibroblasts treated with ethidium bromide and fibroblasts from a patient with mtDNA depletion syndrome. *Biochimica Et Biophysica Acta - Molecular Basis of Disease*, **1997**, 1362, 145-59 | 6.9 | 86 |
| 96 | NDUFA4 mutations underlie dysfunction of a cytochrome c oxidase subunit linked to human neurological disease. *Cell Reports*, **2013**, 3, 1795-805 | 10.6 | 85 |
| 95 | NDUFA4 Mutations Underlie Dysfunction of a Cytochrome c Oxidase Subunit Linked to Human Neurological Disease. *Cell Reports*, **2013**, 4, 402 | 10.6 | 78 |
| 94 | Replication of mitochondrial DNA occurs throughout the mitochondria of cultured human cells. *Experimental Cell Research*, **2003**, 289, 133-42 | 4.2 | 76 |
| 93 | FOXRED1, encoding an FAD-dependent oxidoreductase complex-I-specific molecular chaperone, is mutated in infantile-onset mitochondrial encephalopathy. *Human Molecular Genetics*, **2010**, 19, 4837-47 | 5.6 | 73 |
| 92 | Mitochondrial DNA depletion can be prevented by dGMP and dAMP supplementation in a resting culture of deoxyguanosine kinase-deficient fibroblasts. *Human Molecular Genetics*, **2003**, 12, 1839-45 | 5.6 | 71 |
| 91 | Clinical, biochemical and morphological features of hepatocerebral syndrome with mitochondrial DNA depletion due to deoxyguanosine kinase deficiency. *Journal of Hepatology*, **2005**, 43, 333-41 | 13.4 | 69 |
| 90 | Human cytochrome c oxidase: structure, function, and deficiency. *Journal of Bioenergetics and Biomembranes*, **1997**, 29, 151-63 | 3.7 | 68 |
| 89 | Pathogenic Parkinson's disease mutations across the functional domains of LRRK2 alter the autophagic/lysosomal response to starvation. *Biochemical and Biophysical Research Communications* , **2013**, 441, 862-6 | 3.4 | 66 |
| 88 | Decreased brain protein levels of cytochrome oxidase subunits in Alzheimer's disease and in hereditary spinocerebellar ataxia disorders: a nonspecific change?. *Journal of Neurochemistry*, **1999**, 72, 700-7 | 6 | 62 |
| 87 | Signal transducer and activator of transcription 2 deficiency is a novel disorder of mitochondrial fission. *Brain*, **2015**, 138, 2834-46 | 11.2 | 59 |
| 86 | HIBCH mutations can cause Leigh-like disease with combined deficiency of multiple mitochondrial respiratory chain enzymes and pyruvate dehydrogenase. *Orphanet Journal of Rare Diseases*, **2013**, 8, 188 | 4.2 | 57 |
| 85 | Mutations in the mitochondrial complex I assembly factor NDUFAF1 cause fatal infantile hypertrophic cardiomyopathy. *Journal of Medical Genetics*, **2011**, 48, 691-7 | 5.8 | 55 |
| 84 | Regulation of cytochrome c oxidase by interaction of ATP at two binding sites, one on subunit VIa. *Biochemistry*, **1994**, 33, 11833-41 | 3.2 | 51 |
| 83 | Mitochondrial single-stranded DNA binding protein is required for maintenance of mitochondrial DNA and 7S DNA but is not required for mitochondrial nucleoid organisation. *Biochimica Et Biophysica Acta - Molecular Cell Research*, **2010**, 1803, 931-9 | 4.9 | 50 |

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|---|---|---|---|
| 82 | TRNT1 deficiency: clinical, biochemical and molecular genetic features. *Orphanet Journal of Rare Diseases*, **2016**, 11, 90 | 4.2 | 50 |
| 81 | A LON-ClpP Proteolytic Axis Degrades Complex I to Extinguish ROS Production in Depolarized Mitochondria. *Cell Reports*, **2016**, 17, 2522-2531 | 10.6 | 49 |
| 80 | Influence of mitochondrial DNA level on cellular energy metabolism: implications for mitochondrial diseases. *Journal of Bioenergetics and Biomembranes*, **2008**, 40, 59-67 | 3.7 | 48 |
| 79 | Altered gene expression and functions of mitochondria in human nephrotic syndrome. *FASEB Journal*, **1999**, 13, 523-32 | 0.9 | 47 |
| 78 | Analysis of mutant DNA polymerase gamma in patients with mitochondrial DNA depletion. *Human Mutation*, **2009**, 30, 248-54 | 4.7 | 46 |
| 77 | Mitochondrial DNA depletion syndrome is expressed in amniotic fluid cell cultures. *American Journal of Pathology*, **1999**, 155, 67-70 | 5.8 | 45 |
| 76 | Regulation of the expression of mitochondrial proteins: relationship between mtDNA copy number and cytochrome-c oxidase activity in human cells and tissues. *Biochimica Et Biophysica Acta - Bioenergetics*, **1993**, 1144, 177-83 | 4.6 | 45 |
| 75 | A Human Neural Crest Stem Cell-Derived Dopaminergic Neuronal Model Recapitulates Biochemical Abnormalities in GBA1 Mutation Carriers. *Stem Cell Reports*, **2017**, 8, 728-742 | 8 | 42 |
| 74 | Depletion of mitochondrial DNA in the liver of an infant with neonatal giant cell hepatitis. *Human Pathology*, **2002**, 33, 247-53 | 3.7 | 42 |
| 73 | Somatic copy number gains of □synuclein (SNCA) in Parkinson's disease and multiple system atrophy brains. *Brain*, **2018**, 141, 2419-2431 | 11.2 | 41 |
| 72 | Kearns-Sayre syndrome caused by defective R1/p53R2 assembly. *Journal of Medical Genetics*, **2011**, 48, 610-7 | 5.8 | 41 |
| 71 | Mutant torsinA, which causes early-onset primary torsion dystonia, is redistributed to membranous structures enriched in vesicular monoamine transporter in cultured human SH-SY5Y cells. *Movement Disorders*, **2005**, 20, 432-440 | 7 | 41 |
| 70 | Subunit specific monoclonal antibodies show different steady-state levels of various cytochrome-c oxidase subunits in chronic progressive external ophthalmoplegia. *Biochimica Et Biophysica Acta - Molecular Basis of Disease*, **1996**, 1315, 199-207 | 6.9 | 41 |
| 69 | A novel mutation in the deoxyguanosine kinase gene causing depletion of mitochondrial DNA. *Annals of Neurology*, **2002**, 52, 237-9 | 9.4 | 39 |
| 68 | Steady-state transcript levels of cytochrome c oxidase genes during human myogenesis indicate subunit switching of subunit VIa and co-expression of subunit VIIa isoforms. *Biochimica Et Biophysica Acta - Molecular Basis of Disease*, **1992**, 1139, 155-62 | 6.9 | 39 |
| 67 | Myoclonus-dystonia syndrome with severe depression is caused by an exon-skipping mutation in the epsilon-sarcoglycan gene. *Movement Disorders*, **2007**, 22, 1173-5 | 7 | 38 |
| 66 | Phenotypic variability of mitochondrial disease caused by a nuclear mutation in complex II. *Molecular Genetics and Metabolism*, **2006**, 89, 214-21 | 3.7 | 38 |
| 65 | Creation of an open-access, mutation-defined fibroblast resource for neurological disease research. *PLoS ONE*, **2012**, 7, e43099 | 3.7 | 35 |

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|---|---|---|---|
| 64 | Analysis of the trinucleotide CAG repeat from the DNA polymerase gamma gene (POLG) in patients with Parkinson's disease. *Neuroscience Letters*, **2005**, 376, 56-9 | 3.3 | 35 |
| 63 | Kinetic properties of mutant deoxyguanosine kinase in a case of reversible hepatic mtDNA depletion. *Biochemical Journal*, **2007**, 402, 377-85 | 3.8 | 34 |
| 62 | Odour-conditioned anemotaxis of apterous aphids (Cryptomyzus korschelti) in response to host plants. *Physiological Entomology*, **1987**, 12, 473-479 | 1.9 | 34 |
| 61 | Collated mutations in mitochondrial DNA (mtDNA) depletion syndrome (excluding the mitochondrial gamma polymerase, POLG1). *Biochimica Et Biophysica Acta - Molecular Basis of Disease*, **2009**, 1792, 1109-12 | 6.9 | 33 |
| 60 | Type I tyrosinemia: lack of immunologically detectable fumarylacetoacetase enzyme protein in tissues and cell extracts. *Pediatric Research*, **1987**, 22, 394-8 | 3.2 | 33 |
| 59 | A Clinical, Neuropathological and Genetic Study of Homozygous A467T POLG-Related Mitochondrial Disease. *PLoS ONE*, **2016**, 11, e0145500 | 3.7 | 33 |
| 58 | The clinical spectrum and natural history of early-onset diseases due to DNA polymerase gamma mutations. *Genetics in Medicine*, **2017**, 19, 1217-1225 | 8.1 | 32 |
| 57 | Mitochondria as target to inhibit proliferation and induce apoptosis of cancer cells: the effects of doxycycline and gemcitabine. *Scientific Reports*, **2020**, 10, 4363 | 4.9 | 31 |
| 56 | Analysis of COX2 mutants reveals cytochrome oxidase subassemblies in yeast. *Biochemical Journal*, **2005**, 390, 703-8 | 3.8 | 29 |
| 55 | Assembly of cytochrome c oxidase: what can we learn from patients with cytochrome c oxidase deficiency?. *Biochemical Society Transactions*, **2001**, 29, 446-51 | 5.1 | 29 |
| 54 | NDUFA4 (Renamed COXFA4) Is a Cytochrome-c Oxidase Subunit. *Trends in Endocrinology and Metabolism*, **2018**, 29, 452-454 | 8.8 | 27 |
| 53 | Pathogenic LRRK2 mutations do not alter gene expression in cell model systems or human brain tissue. *PLoS ONE*, **2011**, 6, e22489 | 3.7 | 27 |
| 52 | COX10 mutations resulting in complex multisystem mitochondrial disease that remains stable into adulthood. *JAMA Neurology*, **2013**, 70, 1556-61 | 17.2 | 24 |
| 51 | Characterization of a novel TYMP splice site mutation associated with mitochondrial neurogastrointestinal encephalomyopathy (MNGIE). *Neuromuscular Disorders*, **2009**, 19, 151-4 | 2.9 | 24 |
| 50 | Mitochondrial dysfunction in congenital nephrotic syndrome. *Laboratory Investigation*, **2000**, 80, 1227-32 | 5.9 | 24 |
| 49 | Biochemical, genetic and immunoblot analyses of 17 patients with an isolated cytochrome c oxidase deficiency. *Biochimica Et Biophysica Acta - Molecular Basis of Disease*, **1999**, 1455, 35-44 | 6.9 | 24 |
| 48 | Nucleotide sequence of cDNA encoding human fumarylacetoacetase. *Nucleic Acids Research*, **1990**, 18, 1887 | 20.1 | 24 |
| 47 | Diagnostic value of succinate ubiquinone reductase activity in the identification of patients with mitochondrial DNA depletion. *Journal of Inherited Metabolic Disease*, **2002**, 25, 7-16 | 5.4 | 23 |

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| 46 | Nucleotide sequence of cDNA encoding subunit VIb of human cytochrome c oxidase. *Nucleic Acids Research*, **1989**, 17, 1766 | 20.1 | 22 |
| 45 | A novel mutation in SURF1 causes skipping of exon 8 in a patient with cytochrome c oxidase-deficient leigh syndrome and hypertrichosis. *Molecular Genetics and Metabolism*, **2001**, 73, 340-3 | 3.7 | 20 |
| 44 | DNA isolation protocol effects on nuclear DNA analysis by microarrays, droplet digital PCR, and whole genome sequencing, and on mitochondrial DNA copy number estimation. *PLoS ONE*, **2017**, 12, e0180467 | 3.7 | 18 |
| 43 | Mutations of cytochrome c oxidase subunits 1 and 3 in Saccharomyces cerevisiae: assembly defect and compensation. *Biochimica Et Biophysica Acta - Bioenergetics*, **2002**, 1554, 101-7 | 4.6 | 18 |
| 42 | Mitochondrial cristae remodelling is associated with disrupted OPA1 oligomerisation in the Huntington's disease R6/2 fragment model. *Experimental Neurology*, **2017**, 288, 167-175 | 5.7 | 17 |
| 41 | Assignment of the gene coding for human cytochrome c oxidase subunit VIb to chromosome 19, band q13.1, by fluorescence in situ hybridisation. *Human Genetics*, **1991**, 87, 325-7 | 6.3 | 17 |
| 40 | Selective striatal mtDNA depletion in end-stage Huntington's disease R6/2 mice. *Experimental Neurology*, **2015**, 266, 22-9 | 5.7 | 16 |
| 39 | Immunological phenotyping of fibroblast cultures from patients with a mitochondrial respiratory chain deficit. *Laboratory Investigation*, **2001**, 81, 1069-77 | 5.9 | 15 |
| 38 | Isolation of cDNAs encoding subunit VIb of cytochrome c oxidase and steady-state levels of coxVIb mRNA in different tissues. *Gene*, **1990**, 93, 285-91 | 3.8 | 15 |
| 37 | Clinicopathologic and molecular spectrum of -related mitochondrial disease. *Neurology: Genetics*, **2017**, 3, e149 | 3.8 | 14 |
| 36 | Does doxycycline work in synergy with cisplatin and oxaliplatin in colorectal cancer?. *World Journal of Surgical Oncology*, **2009**, 7, 2 | 3.4 | 14 |
| 35 | Mitochondria as oncotarget: a comparison between the tetracycline analogs doxycycline and COL-3. *Oncotarget*, **2018**, 9, 33818-33831 | 3.3 | 13 |
| 34 | Clonal expansion of T cells in abdominal aortic aneurysm: a role for doxycycline as drug of choice?. *International Journal of Molecular Sciences*, **2015**, 16, 11178-95 | 6.3 | 12 |
| 33 | Increased sensitivity of myoblasts to oxidative stress in amyotrophic lateral sclerosis peripheral tissues. *Experimental Neurology*, **2009**, 218, 92-7 | 5.7 | 12 |
| 32 | Nucleotide sequence of the last exon of the gene for human cytochrome c oxidase subunit VIb and its flanking regions. *Biochimica Et Biophysica Acta Gene Regulatory Mechanisms*, **1991**, 1089, 283-5 | | 10 |
| 31 | Huntingtin Aggregates and Mitochondrial Pathology in Skeletal Muscle but not Heart of Late-Stage R6/2 Mice. *Journal of Huntington's Disease*, **2019**, 8, 145-159 | 1.9 | 9 |
| 30 | Influence of zinc and zinc chelator on HT-29 colorectal cell line. *BioMetals*, **2011**, 24, 143-51 | 3.4 | 9 |
| 29 | Lowering the apoptotic threshold in colorectal cancer cells by targeting mitochondria. *Cancer Cell International*, **2010**, 10, 31 | 6.4 | 9 |

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| 28 | SCID mice containing muscle with human mitochondrial DNA mutations. An animal model for mitochondrial DNA defects. *Journal of Clinical Investigation*, **1998**, 102, 2090-5 | 15.9 | 8 |
| 27 | Expression of Nodulin Genes During Nodule Development from Effective and Ineffective Root Nodules **1984**, 579-586 | | 8 |
| 26 | Intracellular oxygenation and cytochrome oxidase C activity in ischemic preconditioning of steatotic rabbit liver. *American Journal of Surgery*, **2010**, 200, 507-18 | 2.7 | 7 |
| 25 | Identification of three human pseudogenes for subunit VIb of cytochrome c oxidase: a molecular record of gene evolution. *Gene*, **1991**, 102, 237-44 | 3.8 | 7 |
| 24 | Glucosylceramidase degradation in fibroblasts carrying bi-allelic Parkin mutations. *Molecular Genetics and Metabolism*, **2013**, 109, 402-3 | 3.7 | 5 |
| 23 | Mitofusin 1 and mitofusin 2 are ubiquitinated in a PINK1/parkin-dependent manner upon induction of mitophagy. *Human Molecular Genetics*, **2013**, 22, 1697-1697 | 5.6 | 4 |
| 22 | Relapsing neuropathy in an 18-year-old woman. *Lancet Neurology, The*, **2007**, 6, 192-8 | 24.1 | 4 |
| 21 | Sirtuin 5 depletion impairs mitochondrial function in human proximal tubular epithelial cells. *Scientific Reports*, **2021**, 11, 15510 | 4.9 | 3 |
| 20 | Comment on "aneurysmal lesions of patients with abdominal aortic aneurysm contain clonally expanded T cells". *Journal of Immunology*, **2014**, 193, 2041 | 5.3 | 2 |
| 19 | The diagnosis of inherited metabolic diseases by microarray gene expression profiling. *Orphanet Journal of Rare Diseases*, **2010**, 5, 34 | 4.2 | 2 |
| 18 | Measurement of kinetic parameters of human platelet DNA polymerase gamma. *Methods*, **2010**, 51, 374-86 | 4 | 2 |
| 17 | Cytochrome-c Oxidase (COX) | | 2 |
| 16 | Ambroxol reverses tau and □synuclein accumulation in a cholinergic N370S GBA1 mutation model.. *Human Molecular Genetics*, **2022**, | 5.6 | 2 |
| 15 | Subunit composition of respiratory chain complex 1 and its responses to oxygen in mitochondria from human donor livers. *BMC Research Notes*, **2017**, 10, 547 | 2.3 | 1 |
| 14 | Loss of PINK1 or Parkin Function Results in a Progressive Loss of Mitochondrial Function **2017**, 187-209 | | 1 |
| 13 | Chapter 1 Structure and Function of the Mitochondrial Oxidative Phosphorylation System. *Blue Books of Practical Neurology*, **2002**, 1-34 | | 1 |
| 12 | The Human Mitochondrial Genome. *Oxidative Stress and Disease*, **2005**, 95-246 | | 1 |
| 11 | The PINK1 □Parkin mitophagy signalling pathway is not functional in peripheral blood mononuclear cells | | 1 |

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| 10 | The PINK1-Parkin mitophagy signalling pathway is not functional in peripheral blood mononuclear cells. *PLoS ONE*, **2021**, 16, e0259903 | 3.7 | 0 |
| 9 | Mitochondrial DNA: Structure, Genetics, Replication and Defects **2019**, 127-152 | | |
| 8 | B25 Mitochondrial fission and fusion in skeletal muscle from HD patients and zQ175 mice. *Journal of Neurology, Neurosurgery and Psychiatry*, **2016**, 87, A17.3-A18 | 5.5 | |
| 7 | B34 Mitochondrial Biogenesis, And Respiratory Chain Assembly And Function, In Heart Of R6/2 Mouse Model Of Huntington's Disease. *Journal of Neurology, Neurosurgery and Psychiatry*, **2014**, 85, A21-A21 | 5.5 | |
| 6 | D03 Mitochondrial Biogenesis, And Respiratory Chain Assembly And Function, In Skeletal Muscle Of The R6/2 Mouse Model And Human Huntington's Disease. *Journal of Neurology, Neurosurgery and Psychiatry*, **2014**, 85, A32-A32 | 5.5 | |
| 5 | POG01 Anti-thymidine phosphorylase antibodies in the diagnosis of mitochondrial neurogastrointestinal encephalomyopathy. *Journal of Neurology, Neurosurgery and Psychiatry*, **2010**, 81, e48-e48 | 5.5 | |
| 4 | 14 Mitochondrial cardiomyopathy caused by defective assembly of respiratory chain complex I. *Heart*, **2011**, 97, e8-e8 | 5.1 | |
| 3 | Assessment of the significance of mitochondrial DNA damage by chemotherapeutic agents **2005**, 27, 337 | | |
| 2 | Mitochondrial respiratory chain and Krebs cycle enzyme function in human donor livers subjected to end-ischaemic hypothermic machine perfusion. *PLoS ONE*, **2021**, 16, e0257783 | 3.7 | |
| 1 | Mitochondrial DNA Mutations and Nuclear Mitochondrial Interactions in Human Disease **1999**, 635-663 | | |