

LETTER TO THE EDITOR

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# Sorting of exclusive mitochondrial proteins from the Mitoproteome database

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## Abstract

World Wide Web and scientific literature record information regarding several organelles and mitochondria is not an exception. However, currently mitochondrial databases also provide information about non-mitochondrial proteins. As on date, there is no way to know the exclusive mitochondrial proteins. It is crucial to know the exclusively mitochondrial proteins, present in mitochondria but absent elsewhere. In this work, by analysis of available databases, we have provided a list of exclusively mitochondrial proteins known as on date.

**Keywords:** Mitochondrial protein databases, Mitoproteome, Mitochondrial proteins annotation, Sorting exclusive mitochondrial proteins, Mitochondrial biogenesis

## Background

Mitochondria research is a pivot of current research in life sciences. It is unraveling several facts which revolutionize humans understanding about biology. At present, several databases are available dealing functional annotation of proteins and genes with respect to organelle including mitochondria.

Localization of mitochondrial proteins is annotated already (Rath et al. 2021). It has been demonstrated that 99% mitochondrial proteins are nuclear genome encoded. There are only 13 proteins in the mitochondria that are encoded by mitochondrial genome. Different guiding factors like mitochondrial targeting sequence (MTS), translocase of outer membrane (TOM) complex, etc., are established for mitochondrial protein translocation (Nicolas et al. 2019). Another study has reported that hydrophobic membrane proteins encoded by mitochondrial genome are synthesized in endoplasmic reticulum, while nuclear-encoded proteins are synthesized in cytoplasm (Bjorkholm et al. 2015). So, coordination between mitochondrial and nuclear genomes is necessary for

mitochondrial biogenesis at normal and stressed conditions (Lionaki et al. 2016). Very recently, an RNA-binding protein, Clustered mitochondria homolog (CLUH) is identified as mitochondrial protein translation regulator in cytoplasm (Hemono et al. 2022).

Mitochondrial proteins or genes perform numerous and diverse functions in apoptosis, energy conversation, heat production, homeostasis, cancer, ageing, cell signaling and death pathways (Pfanter et al. 2019; Lin et al. 2022). Therefore, mitochondrial protein/gene sequence databases and annotation system are crucial for mitochondrial researchers. Some databases record details about proteins location, function, etc., where mitochondrial proteins are present (The Human Protein Atlas 2022; Helix Mitochondrial DNA Database—Available for Research Analysis-Helix Population Genomics 2022; Organelle Genome Resources 2022). However, it is difficult to figure out which proteins are exclusive mitochondrial and which are not. In this context, we have sorted exclusively mitochondrial proteins from existing database (MitoProteome, OMIM, UniProt, The Human Protein Atlas).

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**Table 1** List of exclusive mitochondrial proteins

| Sr. no | MITO ID  | Gene/protein name   | Source   |
|--------|----------|---|--|
| 1      | MT000648 | A kinase (PRKA) anchor protein 1  | NCBI Gene ID: 8165 & UniProtKB–Q92667 (AKAP1_HUMAN) & PMID 24,412,345  |
| 2      | MT000005 | Acetyl-CoA carboxylase $\beta$  | Gene ID: 32 & UniProtKB–O00763 (ACACB_HUMAN) & <a href="https://doi.org/10.1073/pnas.97.4.1444">https://doi.org/10.1073/pnas.97.4.1444</a>                       |
| 3      | MT000778 | Acetyl-CoA acyltransferase 2  | Gene ID: 10,449 & UniProtKB–P42765 (THIM_HUMAN)  |
| 4      | MT000070 | BCL2-antagonist/killer 1  | Gene ID: 578 & UniProtKB–Q16611 (BAK_HUMAN)  |
| 5      | MT000960 | BCL2-binding component 3  | Gene ID: 27,113 & UniProtKB–Q9BXH1 (BBC3_HUMAN)  |
| 6      | MT000146 | Carnitine palmitoyltransferase 1A (liver)   | Gene ID: 1374 & UniProtKB–P50416 (CPT1A_HUMAN)   |
| 7      | MT001561 | Glycerol-3-phosphate acyltransferase 2, mitochondrial                             | Gene ID: 150,763 & UniProtKB–Q6NUI2 (GPAT2_HUMAN) & <a href="https://doi.org/10.1038/s41387-018-0045-x">https://doi.org/10.1038/s41387-018-0045-x</a>            |
| 8      | MT000664 | Kynurenine 3-monooxygenase (kynurenine 3-hydroxylase)                             | Gene ID: 8564 & UniProtKB–O15229 (KMO_HUMAN) & <a href="https://doi.org/10.1038/cddis.2016.87">https://doi.org/10.1038/cddis.2016.87</a>                         |
| 9      | MT000343 | Monoamine oxidase B   | UniProtKB P27338(AOFB_HUMAN)<br>PMID: 12,379,239; Gene ID: 4129  |
| 10     | MT001517 | Mitochondrial elongation factor 2   | Gene ID: 125,170 & UniProtKB–Q96C03 (MID49_HUMAN)  |
| 11     | MT000800 | Metaxin 2   | Gene ID: 10,651; PMID: 10,381,257 UniProtKB O75431 (MTX2_HUMAN)  |
| 12     | MT001642 | Metaxin 3   | Gene ID: 345,778; PMID: 9,045,676 UniProtKB-Q5HY17 (MTX3_HUMAN)  |
| 13     | MT001315 | NLR family member X1  | Gene ID: 79,671 & UniProtKB–Q86UT6 (NLRX1_HUMAN)<br><a href="https://doi.org/10.1038/nature06501">https://doi.org/10.1038/nature06501</a>                        |
| 14     | MT001589 | Phospholipase D family, member 6  | Gene ID: 201,164 & UniProtKB–Q8N2A8 (PLD6_HUMAN)<br>PMID: <a href="https://pubmed.ncbi.nlm.nih.gov/25926691/">25,926,691</a>                                     |
| 15     | MT000482 | Phorbol-12-myristate-13-acetate-induced protein 1                                 | Gene ID: 5366 & UniProtKB–Q13794 (APR_HUMAN)   |
| 16     | MT001422 | Mitochondria-localized glutamic acid-rich protein                                 | UniProtKB–Q8TDB4 (HUMMR_HUMAN) &   |
| 17     | MT001551 | BRI3-binding protein  | UniProtKB–Q8WY22 (BRI3B_HUMAN) & PMID: 17,943,721  |
| 18     | MT000907 | FK506-binding protein 8, 38 kDa   | UniProtKB–Q14318 (FKBP8_HUMAN)   |
| 19     | MT001549 | FUN14 domain containing 1   | UniProtKB–Q8IVP5 (FUND1_HUMAN)   |
| 20     | MT000342 | Monoamine oxidase A   | UniProtKB–P21397 (AOFA_HUMAN)  |
| 21     | MT000737 | Mitofusin 2   | UniProtKB–O95140 (MFN2_HUMAN) & PubMed: <a href="https://pubmed.ncbi.nlm.nih.gov/11181170/">11,181,170</a>   |
| 22     | MT001151 | Ras homolog family member T1  | UniProtKB–Q8IXI2 (MIRO1_HUMAN) & PMID: 19,528,298  |
| 23     | MT001448 | Ras homolog family member T2  | UniProtKB–Q8IXI1 (MIRO2_HUMAN)   |
| 24     | MT001655 | Translocase of outer mitochondrial membrane 20 homolog (yeast)-like               | UniProtKB–Q6UXN7 (TO20L_HUMAN)   |
| 25     | MT001424 | Ubiquitin specific peptidase 30   | UniProtKB–Q70CQ3 (UBP30_HUMAN)   |
| 26     | MT000626 | Voltage-dependent anion channel 2   | UniProtKB–P45880 (VDAC2_HUMAN)   |
| 27     | MT001581 | Phosphoglycerate mutase family member 5   | UniProtKB–Q96HS1 (PGAM5_HUMAN) & PMID: 18,387,606  |
| 28     | MT000173 | Cytochrome P450, family 27, subfamily B, polypeptide 1                            | UniProtKB–O15528 (CP27B_HUMAN)   |
| 29     | MT000554 | Succinate dehydrogenase complex, subunit D, integral membrane protein             | UniProtKB–O14521 (DHSD_HUMAN)  |
| 30     | MT000641 | Hydroxysteroid (17- $\beta$ ) dehydrogenase 8                                     | UniProtKB–Q92506 (DHB8_HUMAN)  |
| 31     | MT000736 | Translocase of outer mitochondrial membrane 70 homolog A ( <i>S. cerevisiae</i> ) | UniProtKB–O94826 (TOM70_HUMAN)   |
| 32     | MT000779 | Translocase of outer mitochondrial membrane 40 homolog (yeast)                    | UniProtKB–O96008 (TOM40_HUMAN)   |
| 33     | MT000918 | LETM1 domain containing 1   | UniProtKB–Q6P1Q0 (LTMD1_HUMAN)   |
| 34     | MT001391 | Translocase of outer mitochondrial membrane 40 homolog (yeast)-like               | UniProtKB–Q969M1 (TM40L_HUMAN)   |
| 35     | MT001597 | Spermatogenesis-associated 19   | UniProtKB–Q7Z5L4 (SPT19_HUMAN)   |
| 36     | MT000144 | Coproporphyrinogen oxidase  | Gene ID: 1371 & UniProtKB–P36551 (HEM6_HUMAN)<br><a href="https://doi.org/10.1016/b978-012095440-7/50031-7">https://doi.org/10.1016/b978-012095440-7/50031-7</a> |
| 37     | MT000412 | NADH dehydrogenase (ubiquinone) 1 $\beta$ subcomplex, 7, 18 kDa                   | Gene ID: 4713 & UniProtKB–P17568 (NDUB7_HUMAN)   |

**Table 1** (continued)

| Sr. no | MITO ID  | Gene/protein name  | Source   |
|--------|----------|--|--|
| 38     | MT000751 | Peptidylprolyl isomerase F   | Gene ID: 10,105 & UniProtKB–P30405 (PPIF_HUMAN)<br>PMID: 10,406,942  |
| 39     | MT000581 | Steroidogenic acute regulatory protein   | Gene ID: 6770 & UniProtKB–P49675 (STAR_HUMAN)<br><a href="https://doi.org/10.1073/pnas.93.24.13731">https://doi.org/10.1073/pnas.93.24.13731</a> |
| 40     | MT001408 | Apoptogenic 1, mitochondrial IMM   | UniProtKB–Q96IL0 (COA8_HUMAN)  |
| 41     | MT001533 | Coiled-coil-helix-coiled-coil-helix domain containing 4 Mito inter memb. space         | UniProtKB–Q8N4Q1 (MIA40_HUMAN)   |
| 42     | MT000972 | HtrA serine peptidase 2 MIM  | UniProtKB–O43464 (HTRA2_HUMAN)   |
| 43     | MT000773 | Mitochondrial calcium uptake 1 MIM   | UniProtKB–Q9BPX6 (MICU1_HUMAN)   |
| 44     | MT001600 | Mitochondrial calcium uptake 2 MIM   | UniProtKB–Q8IYU8 (MICU2_HUMAN)   |
| 45     | MT000418 | NADH dehydrogenase (ubiquinone) Fe-S protein 1, 75 kDa (NADH-coenzyme Q reductase) MIM | UniProtKB–P28331 (NDU51_HUMAN)   |
| 46     | MT000448 | Optic atrophy 1 (autosomal dominant) MIM   | UniProtKB–O60313 (OPA1_HUMAN)  |
| 47     | MT000947 | Family with sequence similarity 162, member A  | UniProtKB–Q96A26 (F162A_HUMAN)   |
| 48     | MT001404 | Ubiquinol-cytochrome c reductase complex assembly factor 2                             | UniProtKB–Q9BRT2 (UQCC2_HUMAN)   |
| 49     | MT000002 | ATP-binding cassette, subfamily B (MDR/TAP), member 7                                  | UniProtKB–O75027 (ABCB7_HUMAN)   |
| 50     | MT000843 | ATP-binding cassette, subfamily B (MDR/TAP), member 8                                  | UniProtKB–Q9NUT2 (MITOS_HUMAN)   |
| 51     | MT001559 | Apoptosis-inducing factor, mitochondrion-associated, 3                                 | UniProtKB–Q96NN9 (AIFM3_HUMAN)   |
| 52     | MT000526 | Aldehyde dehydrogenase 18 family, member A1  | UniProtKB–P54886 (P5CS_HUMAN)  |
| 53     | MT001548 | Apolipoprotein O-like  | UniProtKB–Q6UXV4 (MIC27_HUMAN)   |
| 54     | MT001145 | ATPase family, AAA domain containing 3A  | UniProtKB–Q9NVI7 (ATD3A_HUMAN)   |
| 55     | MT001384 | ATPase family, AAA domain containing 3B  | UniProtKB–Q5T9A4 (ATD3B_HUMAN)   |
| 56     | MT000081 | 3-Hydroxybutyrate dehydrogenase, type 1  | UniProtKB–Q02338 (BDH_HUMAN)   |
| 57     | MT001121 | Coiled-coil domain containing 109B   | UniProtKB–Q9NWR8 (MCUB_HUMAN)  |
| 58     | MT001405 | Coiled-coil-helix-coiled-coil-helix domain containing 6                                | UniProtKB–Q9BRQ6 (MIC25_HUMAN)   |
| 58     | MT001158 | Choline dehydrogenase  | UniProtKB–Q8NE62 (CHDH_HUMAN)  |
| 60     | MT001679 | Creatine kinase, mitochondrial 1A  | UniProtKB–P12532 (KCRCU_HUMAN)   |
| 61     | MT000120 | Creatine kinase, mitochondrial 2 (sarcomeric)  | UniProtKB–P17540 (KCRS_HUMAN)  |
| 62     | MT000966 | Coenzyme Q2 4-hydroxybenzoate polyprenyltransferase                                    | UniProtKB–Q96H96 (COQ2_HUMAN)  |
| 63     | MT001035 | Coenzyme Q4  | UniProtKB–Q9Y3A0 (COQ4_HUMAN)  |
| 64     | MT001206 | Coenzyme Q9  | UniProtKB–O75208 (COQ9_HUMAN)  |
| 65     | MT000141 | COX10 heme A: farnesyltransferase cytochrome c oxidase assembly factor                 | UniProtKB–Q12887 (COX10_HUMAN)   |
| 66     | MT000142 | COX11 cytochrome c oxidase copper chaperone  | UniProtKB–Q9Y6N1 (COX11_HUMAN)   |
| 67     | MT000753 | TNF receptor-associated protein 1  | UniProtKB–Q12931 (TRAP1_HUMAN) (PMID: 23,564,345)  |
| 68     | MT001419 | Cytochrome c oxidase subunit IV isoform 2 (lung)                                       | UniProtKB–Q96KJ9 (COX42_HUMAN)   |
| 69     | MT000705 | Cytochrome c oxidase subunit Va  | UniProtKB–P20674(COX5A_HUMAN)  |
| 70     | MT000130 | Cytochrome c oxidase subunit Vb  | UniProtKB–P10606 (COX5B_HUMAN)   |
| 71     | MT000131 | Cytochrome c oxidase subunit VIa polypeptide 1   | UniProtKB–P12074 (CX6A1_HUMAN)   |
| 72     | MT000132 | Cytochrome c oxidase subunit VIa polypeptide 2   | UniProtKB–Q02221 (CX6A2_HUMAN)   |
| 73     | MT000133 | Cytochrome c oxidase subunit VIb polypeptide 1 (ubiquitous)                            | UniProtKB–P14854 (CX6B1_HUMAN)   |
| 74     | MT000134 | Cytochrome c oxidase subunit VIc   | UniProtKB–P09669 (COX6C_HUMAN)   |
| 75     | MT000135 | Cytochrome c oxidase subunit VIIa polypeptide 1 (muscle)                               | UniProtKB–P24310 (CX7A1_HUMAN)   |
| 76     | MT000805 | YME1-like 1 ATPase   | UniProtKB–Q96TA2(YMEL1_HUMAN)<br>PMID: 10,843,804; PMID: 22,262,461  |
| 77     | MT000699 | Cytochrome c oxidase subunit VIIa polypeptide 2-like                                   | UniProtKB–O14548(COX7R_HUMAN)  |
| 78     | MT000138 | Cytochrome c oxidase subunit VIId  | UniProtKB–P24311 (COX7B_HUMAN)   |
| 79     | MT001093 | Cardiolipin synthase 1   | UniProtKB–Q9UJA2(CRLS1_HUMAN)  |
| 80     | MT000162 | Cytochrome c-1   | UniProtKB–P08574 (CY1_HUMAN)   |
| 81     | MT000958 | Ubiquinol-cytochrome c reductase, complex III subunit VII, 9,5 kDa                     | UniProtKB–O14949 (QCR8_HUMAN)  |

**Table 1** (continued)

| Sr. no | MITO ID  | Gene/protein name   | Source  |
|--------|----------|---|---|
| 82     | MT000182 | Dihydroorotate dehydrogenase (quinone)  | UniProtKB–Q02127 (PYRD_HUMAN)                                       |
| 83     | MT001172 | DnaJ (Hsp40) homolog, subfamily C, member 11  | UniProtKB–Q9NVH1 (DJC11_HUMAN)                                      |
| 84     | MT000990 | DnaJ (Hsp40) homolog, subfamily C, member 15  | UniProtKB–Q9Y5T4 (DJC15_HUMAN)                                      |
| 85     | MT000622 | Ubiquinol-cytochrome c reductase hinge protein  | UniProtKB–P07919 (QCR6_HUMAN)                                       |
| 86     | MT000177 | Enoyl-CoA $\delta$ isomerase 1  | UniProtKB–P42126 (ECI1_HUMAN)                                       |
| 87     | MT000223 | Ferredoxin reductase  | UniProtKB–P22570 (ADRO_HUMAN)                                       |
| 88     | MT000224 | Ferrochelatase  | UniProtKB–P22830 (HEMH_HUMAN)                                       |
| 89     | MT000240 | Glutaryl-CoA dehydrogenase  | UniProtKB–Q92947 (GCDH_HUMAN)                                       |
| 90     | MT000957 | Growth hormone inducible transmembrane protein  | UniProtKB–Q9H3K2 (GHITM_HUMAN)                                      |
| 91     | MT000262 | Glycerol-3-phosphate dehydrogenase 2 (mitochondrial)  | UniProtKB–P43304 (GPDM_HUMAN)                                       |
| 92     | MT000279 | Hydroxyacyl-CoA dehydrogenase   | UniProtKB–Q16836 (HCDH_HUMAN)                                       |
| 93     | MT000277 | Hydroxyacyl-CoA dehydrogenase/3-ketoacyl-CoA thiolase/enoyl-CoA hydratase (trifunctional protein), $\alpha$ subunit | UniProtKB–P40939 (ECHA_HUMAN)                                       |
| 94     | MT000929 | HIG1 hypoxia inducible domain family, member 1A   | UniProtKB–Q9Y241 (HIG1A_HUMAN)                                      |
| 95     | MT001582 | HIG1 hypoxia inducible domain family, member 2A   | UniProtKB–Q9BW72 (HIG2A_HUMAN)                                      |
| 96     | MT000275 | Hydroxysteroid (17- $\beta$ ) dehydrogenase 10  | UniProtKB–Q99714 (HCD2_HUMAN)<br>PMID: 12,917,011 PMID: 18,984,158  |
| 97     | MT000312 | Isocitrate dehydrogenase 2 (NADP <sup>+</sup> ), mitochondrial  | UniProtKB–P48735 (IDHP_HUMAN)                                       |
| 98     | MT000827 | Inner membrane protein, mitochondrial   | UniProtKB–Q16891 (MIC60_HUMAN)                                      |
| 99     | MT001584 | Lactate dehydrogenase D   | UniProtKB–Q86WU2 (LDHD_HUMAN)                                       |
| 100    | MT001068 | Mitochondrial pyruvate carrier 1  | UniProtKB–Q9Y5U8 (MPC1_HUMAN)                                       |
| 101    | MT000917 | Mitochondrial pyruvate carrier 2  | UniProtKB–Q95563 (MPC2_HUMAN)                                       |
| 102    | MT000366 | MpV17 mitochondrial inner membrane protein  | UniProtKB–P39210 (MPV17_HUMAN)<br>PMID: 16,582,907 PMID: 16,582,910 |
| 103    | MT001218 | MRS2 magnesium transporter  | UniProtKB–Q9HD23 (MRS2_HUMAN)                                       |
| 104    | MT001675 | Methylenetetrahydrofolate dehydrogenase (NADP <sup>+</sup> dependent) 2-like  | UniProtKB–Q9H903 (MTD2L_HUMAN)                                      |
| 105    | MT000395 | NADH dehydrogenase (ubiquinone) 1 $\alpha$ subcomplex, 1, 7.5 kDa   | UniProtKB–O15239 (NDUA1_HUMAN)                                      |
| 106    | MT001668 | Solute carrier family 25, member 53   | UniProtKB–Q5H9E4 (S2553_HUMAN)                                      |
| 107    | MT001701 | NDUFC2-KCTD14 readthrough   | UniProtKB E9PQ53 (NDUCR_HUMAN)                                      |
| 108    | MT000421 | NADH dehydrogenase (ubiquinone) flavoprotein 1, 51 kDa  | UniProtKB–P49821 (NDUV1_HUMAN)                                      |
| 109    | MT000434 | NME/NM23 nucleoside diphosphate kinase 4  | UniProtKB–O00746 (NDKM_HUMAN)                                       |
| 110    | MT000894 | Nicotinamide nucleotide transhydrogenase  | UniProtKB–Q13423 (NNTM_HUMAN)                                       |
| 111    | MT001400 | Nitric oxide-associated 1   | UniProtKB–Q8NC60 (NOA1_HUMAN)                                       |
| 112    | MT001491 | OMA1 zinc metallopeptidase  | UniProtKB–Q96E52 (OMA1_HUMAN)                                       |
| 113    | MT000451 | Oxidase (cytochrome c) assembly 1-like  | UniProtKB–Q15070 (OXA1L_HUMAN)                                      |
| 114    | MT000457 | Pyruvate carboxylase  | UniProtKB–P11498 (PYC_HUMAN)  |
| 115    | MT000468 | Pyruvate dehydrogenase kinase, isozyme 4  | UniProtKB–Q16654 (PDK4_HUMAN)                                       |
| 116    | MT000714 | Phosphatidylglycerophosphate synthase 1   | UniProtKB–Q32NB8 (PGPS1_HUMAN)                                      |
| 117    | MT000873 | Peptidase (mitochondrial processing) $\alpha$   | UniProtKB–Q10713 (MPPA_HUMAN); PMID: 25,808,372                     |
| 118    | MT000715 | Peptidase (mitochondrial processing) $\beta$  | UniProtKB–O75439 (MPPB_HUMAN); PMID: 22,354,088                     |
| 119    | MT000484 | Polymerase (DNA directed), $\gamma$   | UniProtKB–P54098 (DPOG1_HUMAN); PMID: 18,063,578                    |
| 120    | MT000964 | PRELI domain containing 1   | UniProtKB–Q9Y255 (PRLD1_HUMAN)                                      |
| 121    | MT000511 | Proline dehydrogenase (oxidase) 1   | UniProtKB–O43272 (PROD_HUMAN)                                       |
| 122    | MT001488 | Protein tyrosine phosphatase, mitochondrial 1   | UniProtKB–Q8WUKO (PTPM1_HUMAN)                                      |
| 123    | MT000621 | Ubiquinol-cytochrome c reductase, Rieske iron sulfur polypeptide 1  | UniProtKB–P47985 (UCRI_HUMAN)                                       |
| 124    | MT000618 | Ubiquinol-cytochrome c reductase-binding protein  | UniProtKB–P14927 (QCR7_HUMAN)                                       |
| 125    | MT000549 | SCO1 cytochrome c oxidase assembly protein  | UniProtKB–O75880 (SCO1_HUMAN); PMID: 15,229,189                     |
| 126    | MT000740 | SCO2 cytochrome c oxidase assembly protein  | UniProtKB–O43819 (SCO2_HUMAN); PMID: 15,229,189                     |
| 127    | MT000551 | Succinate dehydrogenase complex, subunit A, flavoprotein (Fp)   | UniProtKB–P31040 (SDHA_HUMAN)                                       |

**Table 1** (continued)

| Sr. no | MITO ID  | Gene/protein name  | Source  |
|--------|----------|--|---|
| 128    | MT000552 | Succinate dehydrogenase complex, subunit B, iron sulfur (lp)   | UniProtKB–P21912 (SDHB_HUMAN)                                     |
| 129    | MT001478 | sideroflexin 1   | UniProtKB–Q9H9B4 (SFXN1_HUMAN); PMID: 30,442,778                  |
| 130    | MT000564 | Solute carrier family 25 (mitochondrial carrier; citrate transporter), member 1                      | UniProtKB–P53007 (TXTP_HUMAN)                                     |
| 131    | MT000035 | Solute carrier family 25 (mitochondrial carrier; adenine nucleotide translocator), member 5          | UniProtKB–P05141 (ADT2_HUMAN)                                     |
| 132    | MT001465 | Solute carrier family 25, member 51  | UniProtKB–Q9H1U9 (S2551_HUMAN)                                    |
| 133    | MT000587 | surfeit 1  | UniProtKB–Q15526 (SURF1_HUMAN)                                    |
| 134    | MT001534 | TAM41, mitochondrial translocator assembly and maintenance protein, homolog ( <i>S. cerevisiae</i> ) | UniProtKB–Q96BW9(TAM41_HUMAN)                                     |
| 135    | MT001393 | Transmembrane protein 126A   | UniProtKB–Q9H061(T126A_HUMAN); PMID: 19,327,736; PMID: 23,500,070 |
| 136    | MT001109 | Transmembrane protein 70   | UniProtKBQ9BUB7(TMM70_HUMAN); PMID: 20,937,241                    |
| 137    | MT000605 | Thiosulfate sulfurtransferase (rhodanese)  | UniProtKB–Q16762 (THTR_HUMAN)                                     |
| 138    | MT001099 | Tetratricopeptide repeat domain 19   | UniProtKB–Q6DKK2(TTC19_HUMAN) PMID: 21,278,747                    |
| 139    | MT000992 | Ubiquinol-cytochrome c reductase, complex III subunit X  | UniProtKB–Q9UDW1(QCR9_HUMAN)                                      |
| 140    | MT000825 | Ubiquinol-cytochrome c reductase, complex III subunit XI   | UniProtKB–O14957 (QCR10_HUMAN)                                    |
| 141    | MT000053 | ATP synthase, H + transporting, mitochondrial F1 complex, $\delta$ subunit                           | UniProtKB–P30049 (ATPD_HUMAN),                                    |
| 142    | MT000054 | ATP synthase, H + transporting, mitochondrial F1 complex, $\epsilon$ subunit                         | UniProtKB–P56381 (ATP5E_HUMAN),                                   |
| 143    | MT000055 | ATP synthase, H + transporting, mitochondrial Fo complex, subunit B1                                 | UniProtKB–P24539 (AT5F1_HUMAN),                                   |
| 144    | MT000056 | ATP synthase, H + transporting, mitochondrial Fo complex, subunit C1 (subunit 9)                     | UniProtKB–P05496 (AT5G1_HUMAN),                                   |
| 145    | MT000058 | ATP synthase, H + transporting, mitochondrial Fo complex, subunit C3 (subunit 9)                     | UniProtKB–P48201 (AT5G3_HUMAN),                                   |
| 146    | MT000098 | Solute carrier family 25 (carnitine/acylcarnitine translocase), member 20                            | UniProtKB–O43772 (MCAT_HUMAN),                                    |
| 147    | MT000156 | Solute carrier family 25 (mitochondrial carrier; dicarboxylate transporter), member 10               | UniProtKB–Q9UBX3 (DIC_HUMAN),                                     |
| 148    | MT000171 | Cytochrome P450, family 24, subfamily A, polypeptide 1   | UniProtKB–Q07973 (CP24A_HUMAN)                                    |
| 149    | MT000333 | Leucine zipper-EF-hand-containing transmembrane protein 1  | UniProtKB–O95202(LETM1_HUMAN)                                     |
| 150    | MT000407 | NADH dehydrogenase (ubiquinone) 1 $\beta$ subcomplex, 2, 8 kDa                                       | UniProtKB–O95178(NDUB2_HUMAN)                                     |
| 151    | MT000413 | NADH dehydrogenase (ubiquinone) 1 $\beta$ subcomplex, 8, 19 kDa                                      | UniProtKB–O95169(NDUB8_HUMAN)                                     |
| 152    | MT000414 | NADH dehydrogenase (ubiquinone) 1 $\beta$ subcomplex, 9, 22 kDa                                      | UniProtKB Q9Y6M9(NDUB9_HUMAN)                                     |
| 153    | MT000416 | NADH dehydrogenase (ubiquinone) 1, subcomplex unknown, 1, 6 kDa                                      | UniProtKB–O43677(NDUC1_HUMAN)                                     |
| 154    | MT000419 | NADH dehydrogenase (ubiquinone) Fe-S protein 2, 49 kDa (NADH-coenzyme Q reductase)                   | UniProtKB–O75306(NDUS2_HUMAN)                                     |
| 155    | MT000420 | NADH dehydrogenase (ubiquinone) Fe-S protein 3, 30 kDa (NADH-coenzyme Q reductase)                   | UniProtKB–O75489(NDUS3_HUMAN)                                     |
| 156    | MT000426 | NADH dehydrogenase (ubiquinone) flavoprotein 2, 24 kDa   | UniProtKB–P19404(NDUV2_HUMAN)                                     |
| 157    | MT000427 | NADH dehydrogenase (ubiquinone) flavoprotein 3, 10 kDa   | UniProtKB–P56181(NDUV3_HUMAN)                                     |
| 158    | MT000474 | Solute carrier family 25 (mitochondrial carrier; phosphate carrier), member 3                        | UniProtKB–Q00325 (MPCP_HUMAN)                                     |
| 159    | MT000570 | Superoxide dismutase 2, mitochondrial  | UniProtKB–P04179 (SODM_HUMAN)                                     |
| 160    | MT000620 | Ubiquinol-cytochrome c reductase core protein II   | UniProtKB–P22695 (QCR2_HUMAN)                                     |
| 161    | MT000645 | Solute carrier family 25 (mitochondrial carrier), member 16  | UniProtKB–P16260 (GDC_HUMAN)                                      |
| 162    | MT000654 | Solute carrier family 25 (mitochondrial carrier; oxoglutarate carrier), member 11                    | UniProtKB–Q02978 (M2OM_HUMAN)                                     |
| 163    | MT001641 | Cytochrome c oxidase subunit VIIIc   | UniProtKB–Q7Z4L0(COX8C_HUMAN)                                     |

**Table 1** (continued)

| Sr. no | MITO ID  | Gene/protein name   | Source                         |
|--------|----------|---|--------------------------------|
| 164    | MT000948 | Translocase of inner mitochondrial membrane 10 homolog B                        | UniProtKB-Q9Y5J6 (T10B_HUMAN)  |
| 165    | MT001052 | Solute carrier family 25 (mitochondrial iron transporter), member 37            | UniProtKB Q9NYZ2(MFRN1_HUMAN)  |
| 166    | MT001059 | Mitochondrial fission process 1   | UniProtKB Q9UDX5(MTFP1_HUMAN)  |
| 167    | MT001088 | NADH dehydrogenase (ubiquinone) 1 $\beta$ subcomplex, 11, 17.3 kDa              | UniProtKB Q9NX14(NDUBB_HUMAN)  |
| 168    | MT001233 | Sulfide quinone reductase-like  | UniProtKB-Q9Y6N5 (SQOR_HUMAN)  |
| 169    | MT001306 | NADH dehydrogenase (ubiquinone) complex I, assembly factor 5                    | UniProtKB Q5TEU4(NDUF5_HUMAN)  |
| 170    | MT001324 | Solute carrier family 25 (mitochondrial carrier: glutamate), member 22          | UniProtKB-Q9H936 (GHC1_HUMAN)  |
| 171    | MT001336 | L-2-hydroxyglutarate dehydrogenase  | UniProtKB-Q9H9P8(L2HDH_HUMAN)  |
| 172    | MT001474 | Coenzyme Q10 homolog A  | UniProtKB-Q96MF6(CQ10A_HUMAN)  |
| 173    | MT001479 | Sideroflexin 5  | UniProtKB-Q8TD22(SFXN5_HUMAN)  |
| 174    | MT001507 | Sideroflexin 4  | UniProtKB-Q6P4A7(SFXN4_HUMAN)  |
| 175    | MT000809 | Caseinolytic mitochondrial matrix peptidase chaperone subunit                   | UniProtKB-O76031 (CLPX_HUMAN)  |
| 176    | MT000713 | Solute carrier family 25, member 27   | UniProtKB-O95847 (UCP4_HUMAN)  |
| 177    | MT000059 | ATP synthase, H <sup>+</sup> transporting, mitochondrial Fo complex, subunit E  | UniProtKB-P56385 (ATP5L_HUMAN) |
| 178    | MT000060 | ATP synthase, H <sup>+</sup> transporting, mitochondrial Fo complex, subunit F6 | UniProtKB-P18859 (ATP5J_HUMAN) |
| 179    | MT000064 | ATP synthase, H <sup>+</sup> transporting, mitochondrial F1 complex, O subunit  | UniProtKB-P48047 (ATPO_HUMAN)  |
| 180    | MT000143 | Cytochrome c oxidase assembly homolog 15 (yeast)                                | UniProtKB Q7KZN9(COX15_HUMAN)  |
| 181    | MT000396 | NADH dehydrogenase (ubiquinone) 1 $\alpha$ subcomplex, 2, 8 kDa                 | UniProtKB-O43678(NDUA2_HUMAN)  |
| 182    | MT000404 | NADH dehydrogenase (ubiquinone) 1 $\alpha$ subcomplex, 10, 42 kDa               | UniProtKB-O95299(NDUAA_HUMAN)  |
| 183    | MT000415 | NADH dehydrogenase (ubiquinone) 1 $\beta$ subcomplex, 10, 22 kDa                | UniProtKB-O96000(NDUBA_HUMAN)  |
| 184    | MT000560 | Solute carrier family 3 (amino acid transporter heavy chain), member 1          | UniProtKB-Q07837 (SLC31_HUMAN) |
| 185    | MT000666 | Solute carrier family 25 (aspartate/glutamate carrier), member 12               | UniProtKB-O75746 (CMC1_HUMAN)  |
| 186    | MT000720 | ATP synthase, H <sup>+</sup> transporting, mitochondrial Fo complex, subunit F2 | UniProtKB-P56134 (ATPK_HUMAN)  |
| 187    | MT000738 | Endo/exonuclease (5'-3'), endonuclease G-like                                   | UniProtKB-Q9Y2C4 (EXO_G_HUMAN) |
| 188    | MT000783 | ATP synthase, H <sup>+</sup> transporting, mitochondrial Fo complex, subunit d  | UniProtKB-O75947 (ATP5H_HUMAN) |
| 189    | MT000853 | Tumor suppressor candidate 2  | UniProtKB-O75896 (TUSC2_HUMAN) |
| 190    | MT000889 | ATP-binding cassette, subfamily B (MDR/TAP), member 10                          | UniProtKBQ9NRK6 (ABCBA_HUMAN)  |
| 191    | MT000909 | Mitochondrial carrier 1   | UniProtKB Q9NZJ7 (MTCH1_HUMAN) |
| 192    | MT000910 | Mitochondrial carrier 2   | UniProtKB Q9Y6C9 (MTCH2_HUMAN) |
| 193    | MT001672 | Mitochondrial inner membrane organizing system 1                                | UniProtKB-Q5TGZ0 (MIC10_HUMAN) |
| 194    | MT000997 | Solute carrier family 25 (mitochondrial carrier; phosphate carrier), member 24  | UniProtKB-Q6NUK1(SCMC1_HUMAN)  |
| 195    | MT001024 | NADH dehydrogenase (ubiquinone) 1 $\alpha$ subcomplex, 13                       | UniProtKB Q9P0J0 (NDUAD_HUMAN) |
| 196    | MT001030 | NADH dehydrogenase (ubiquinone) complex I, assembly factor 1                    | UniProtKB-Q9Y375 (CIA30_HUMAN) |
| 197    | MT001063 | Solute carrier family 25, member 39   | UniProtKB-Q9BZJ4 (S2539_HUMAN) |
| 198    | MT001111 | Solute carrier family 25, member 38   | UniProtKB-Q96DW6 (S2538_HUMAN) |
| 199    | MT001143 | Solute carrier family 25 (pyrimidine nucleotide carrier), member 36             | UniProtKB-Q96CQ1 (S2536_HUMAN) |
| 200    | MT001187 | Solute carrier family 25, member 40   | UniProtKB-Q8TBP6 (S2540_HUMAN) |

**Table 1** (continued)

| Sr. no | MITO ID  | Gene/protein name   | Source   |
|--------|----------|---|--|
| 201    | MT001242 | GUF1 GTPase homolog ( <i>S. cerevisiae</i> )  | UniProtKB–Q8N442 (GUF1_HUMAN)                                      |
| 202    | MT001244 | Mitochondrial ribosomal protein L17   | UniProtKB–Q9NRX2 (RM17_HUMAN)                                      |
| 203    | MT001249 | Mitochondrial calcium uniporter regulator 1   | UniProtKBQ96AQ8 (MCUR1_HUMAN)                                      |
| 204    | MT001305 | Solute carrier family 25 (mitochondrial carrier; phosphate carrier), member 23      | UniProtKB Q9BV35 (SCMC3_HUMAN)                                     |
| 205    | MT001339 | Solute carrier family 8 (sodium/lithium/calcium exchanger), member B1               | UniProtKB–Q6J4K2 (NCLX_HUMAN)                                      |
| 206    | MT001346 | Coenzyme Q10 homolog B ( <i>S. cerevisiae</i> )                                     | UniProtKB Q9H8M1 (CQ10B_HUMAN)                                     |
| 207    | MT001352 | EF-hand domain family, member D1  | UniProtKB Q9BUP0 (EFHD1_HUMAN)                                     |
| 208    | MT001361 | Solute carrier family 25 (mitochondrial folate carrier), member 32                  | UniProtKB–Q9H2D1 (MFTC_HUMAN)                                      |
| 209    | MT001381 | Solute carrier family 25 (glutamate carrier), member 18                             | UniProtKB–Q9H1K4 (GHC2_HUMAN)                                      |
| 210    | MT001386 | Solute carrier family 25 (mitochondrial carrier; ornithine transporter) member 2    | UniProtKB–Q9BXI2 (ORNT2_HUMAN)                                     |
| 211    | MT001388 | Spinster homolog 1 ( <i>Drosophila</i> )  | UniProtKB–Q9H2V7 (SPNS1_HUMAN)                                     |
| 212    | MT001402 | Solute carrier family 25 (pyrimidine nucleotide carrier), member 33                 | UniProtKB–Q9BSK2 (S2533_HUMAN)                                     |
| 213    | MT001446 | Solute carrier family 25 (mitochondrial oxoadipate carrier), member 21              | UniProtKB–Q9BQT8 (ODC_HUMAN)                                       |
| 214    | MT001453 | Mitochondrial calcium uniporter   | UniProtKB–Q8NE86 (MCU_HUMAN)                                       |
| 215    | MT001458 | Solute carrier family 25, member 46   | UniProtKB–Q96AG3 (S2546_HUMAN)                                     |
| 216    | MT001463 | Single-pass membrane protein with aspartate-rich tail 1                             | UniProtKB–Q9H4I9 (EMRE_HUMAN)                                      |
| 217    | MT001487 | Solute carrier family 25 (mitochondrial carrier; phosphate carrier), member 25      | UniProtKBQ6KCM7 (SCMC2_HUMAN)                                      |
| 218    | MT001492 | Solute carrier family 25 (S-adenosylmethionine carrier), member 26                  | UniProtKB Q70HW3 (SAMC_HUMAN)                                      |
| 219    | MT001511 | Solute carrier family 25 (mitochondrial carnitine/acylcarnitine carrier), member 29 | UniProtKBQ8N8R3 (MCATL_HUMAN)                                      |
| 220    | MT001541 | GrpE-like 2, mitochondrial ( <i>E. coli</i> )                                       | UniProtKB Q8TAA5 (GRPE2_HUMAN)                                     |
| 221    | MT001546 | Leucine zipper-EF-hand containing transmembrane protein 2                           | UniProtKB Q2VYF4 (LETM2_HUMAN)                                     |
| 222    | MT001558 | Solute carrier family 25, member 52   | UniProtKB–Q3SY17 (S2552_HUMAN)                                     |
| 223    | MT001565 | Solute carrier family 25, member 48   | UniProtKB–Q6ZT89 (S2548_HUMAN)                                     |
| 224    | MT001579 | Cytochrome c oxidase subunit VIIIb2   | UniProtKB–Q8TF08 (CX7B2_HUMAN)                                     |
| 225    | MT001594 | Solute carrier family 25, member 43   | UniProtKB–Q8WUT9 (S2543_HUMAN)                                     |
| 226    | MT001605 | Solute carrier family 25, member 30   | UniProtKB Q5SVS4 (KMCP1_HUMAN)                                     |
| 227    | MT001614 | ATP synthase, H <sup>+</sup> transporting, mitochondrial Fo complex, subunit G2     | UniProtKB–Q7Z4Y8 (AT5L2_HUMAN)                                     |
| 228    | MT001617 | Solute carrier family 25, member 45   | UniProtKB–Q8N413 (S2545_HUMAN)                                     |
| 229    | MT001621 | Solute carrier family 25, member 47   | UniProtKB–Q6Q0C1 (S2547_HUMAN)                                     |
| 230    | MT001627 | Solute carrier family 25, member 41   | UniProtKB–Q8N5S1 (S2541_HUMAN)                                     |
| 231    | MT001628 | Solute carrier family 25, member 42   | UniProtKB–Q86VD7 (S2542_HUMAN)                                     |
| 232    | MT001630 | Solute carrier family 25, member 34   | UniProtKB–Q6PIV7 (S2534_HUMAN)                                     |
| 233    | MT001643 | Mitochondrial pyruvate carrier 1-like   | UniProtKB P0DKB6 (MPC1L_HUMAN)                                     |
| 234    | MT001648 | NADH dehydrogenase (ubiquinone) Fe-S protein 7, 20 kDa (NADH-coenzyme Q reductase)  | UniProtKB–O75251 (NDUS7_HUMAN)                                     |
| 235    | MT001661 | Solute carrier family 25, member 35   | UniProtKB–Q3KQZ1 (S2535_HUMAN)                                     |
| 236    | MT000140 | <b>Cytochrome c oxidase subunit VIIIa (ubiquitous)</b>                              | UniProtKB–P10176 (COX8A_HUMAN)                                     |
| 237    | MT000169 | <b>Cytochrome P450, family 11, subfamily B, polypeptide 2</b>                       | UniProtKB–P19099 (C11B2_HUMAN)                                     |
| 238    | MT000760 | <b>Coenzyme Q7 homolog, ubiquinone (yeast)</b>                                      | UniProtKB–Q99807 (COQ7_HUMAN)                                      |
| 239    | MT000755 | Amino adipate-semialdehyde synthase   | UniProtKB–Q9UDR5 (AASS_HUMAN)                                      |
| 240    | MT000001 | 4-Aminobutyrate aminotransferase  | UniProtKB–P80404 (GABT_HUMAN), PMID: 15,528,998                    |
| 241    | MT000954 | Acyl-CoA dehydrogenase family, member 8   | UniProtKB–Q9UKU7 (ACAD8_HUMAN), PMID: 11,013,134; PMID: 12,359,132 |

**Table 1** (continued)

| Sr. no | MITO ID  | Gene/protein name   | Source   |
|--------|----------|---|--|
| 242    | MT000006 | Acyl-CoA dehydrogenase, long chain                              | UniProtKB–P28330 (ACADL_HUMAN)   |
| 243    | MT000007 | Acyl-CoA dehydrogenase, C-4 to C-12 straight chain              | UniProtKB–P11310 (ACADM_HUMAN)   |
| 244    | MT000008 | Acyl-CoA dehydrogenase, C-2 to C-3 short chain                  | UniProtKB–P16219 (ACADS_HUMAN)   |
| 245    | MT000009 | Acyl-CoA dehydrogenase, short/branched chain                    | UniProtKB–P45954 (ACDSB_HUMAN)   |
| 246    | MT000010 | Acyl-CoA dehydrogenase, very long chain                         | UniProtKB–P49748 (ACADV_HUMAN)   |
| 247    | MT000011 | Acetyl-CoA acetyltransferase 1                                  | UniProtKB–P24752 (THIL_HUMAN), PMID: 1,979,337                                       |
| 248    | MT000014 | Aconitase 2, mitochondrial                                      | UniProtKB–Q99798 (ACON_HUMAN)  |
| 249    | MT001497 | Acyl-CoA synthetase medium-chain family member 1                | UniProtKB–Q08AH1 (ACSM1_HUMAN)   |
| 250    | MT001644 | Acyl-CoA synthetase medium-chain family member 2B               | UniProtKB–Q68CK6 (ACS2B_HUMAN), PMID: 12,616,642; PMID: 10,434,065; PMID: 19,634,011 |
| 251    | MT000546 | Acyl-CoA synthetase medium-chain family member 3                | UniProtKB–Q53FZ2 (ACSM3_HUMAN)   |
| 252    | MT001411 | Acyl-CoA synthetase short-chain family member 1                 | UniProtKB–Q9NUB1 (ACS2L_HUMAN), PMID: 16,788,062                                     |
| 253    | MT001545 | alcohol dehydrogenase, iron containing, 1                       | UniProtKB–Q8IWW8 (HOT_HUMAN)   |
| 254    | MT001271 | Alanine-glyoxylate aminotransferase 2                           | UniProtKB–Q9BYV1 (AGT2_HUMAN), PMID: 20,018,850; PMID: 24,586,340                    |
| 255    | MT001005 | Adenylate kinase 3  | UniProtKB–Q9UIJ7 (KAD3_HUMAN), PMID: 11,485,571                                      |
| 256    | MT000020 | Adenylate kinase 4  | UniProtKB–P27144 (KAD4_HUMAN), PMID: 11,485,571; PMID: 19,766,732; PMID: 26,980,435  |
| 257    | MT000022 | 5'-Aminolevulinatase synthase 1                                 | UniProtKB–P13196 (HEM1_HUMAN)  |
| 258    | MT000023 | 5'-Aminolevulinatase synthase 2                                 | UniProtKB–P22557 (HEM0_HUMAN), PMID: 14,643,893                                      |
| 259    | MT000026 | Aldehyde dehydrogenase 2 family (mitochondrial)                 | UniProtKB–P05091 (ALDH2_HUMAN)   |
| 260    | MT001397 | alkB, alkylation repair homolog 7 (E. coli)                     | UniProtKB–Q9BT30 (ALKB7_HUMAN), PMID: 23,666,923                                     |
| 261    | MT000041 | Arginase 2  | UniProtKB–P78540 (ARGI2_HUMAN), PMID: 8,898,077                                      |
| 262    | MT000066 | AU RNA-binding protein/enoyl-CoA hydratase                      | UniProtKB–Q13825 (AUHM_HUMAN)  |
| 263    | MT000074 | Branched chain keto acid dehydrogenase E1, $\alpha$ polypeptide | UniProtKB–P12694 (ODBA_HUMAN)  |
| 264    | MT000075 | Branched chain keto acid dehydrogenase E1, $\beta$ polypeptide  | UniProtKB–P21953 (ODBB_HUMAN)  |
| 265    | MT000097 | Carbonic anhydrase VA, mitochondrial                            | UniProtKB–P35218 (CAH5A_HUMAN)   |
| 266    | MT000849 | Carbonic anhydrase VB, mitochondrial                            | UniProtKB–Q9Y2D0(CAH5B_HUMAN)  |
| 267    | MT001428 | Carbonyl reductase 4  | UniProtKB–Q8N4T8 (CBR4_HUMAN), PMID: 19,000,905 PMID: 19,571,038                     |
| 268    | MT000649 | Caseinolytic mitochondrial matrix peptidase proteolytic subunit | UniProtKB–Q16740 (CLPP_HUMAN), PMID: 10,525,407 PMID: 22,354,088                     |
| 269    | MT000154 | citrate synthase  | UniProtKB–O75390 (CISY_HUMAN), PMID: 9,543,345                                       |
| 270    | MT000167 | Cytochrome P450, family 11, subfamily A, polypeptide 1          | UniProtKB–P05108 (CP11A_HUMAN)   |
| 271    | MT000172 | Cytochrome P450, family 27, subfamily A, polypeptide 1          | UniProtKB–Q02318 (CP27A_HUMAN)<br>The Human protein ATLAS                            |
| 272    | MT001686 | D-2-hydroxyglutarate dehydrogenase                              | UniProtKB–Q8N465 (D2HDH_HUMAN), PMID: 15,070,399<br>The Human protein ATLAS          |
| 273    | MT000639 | Death-associated protein 3                                      | UniProtKB–P51398 (RT29_HUMAN), PMID: 11,162,496; PMID: 20,563,667                    |
| 274    | MT001138 | Aspartyl-tRNA synthetase 2, mitochondrial                       | UniProtKB–Q6PI48 (SYDM_HUMAN), PMID: 15,779,907                                      |
| 275    | MT000176 | Dihydrolipoamide branched chain transacylase E2                 | UniProtKB–P11182 (ODB2_HUMAN)  |
| 276    | MT000181 | Deoxyguanosine kinase   | UniProtKB–Q16854 (DGUOK_HUMAN)   |
| 277    | MT001587 | Dihydrofolate reductase-like 1                                  | UniProtKB–Q86XF0 (DYR2_HUMAN), PMID: 21,876,188                                      |
| 278    | MT000185 | Dihydrolipoamide S-acetyltransferase                            | UniProtKB–P10515 (ODP2_HUMAN)  |
| 279    | MT000998 | Dimethylglycine dehydrogenase                                   | UniProtKB–Q9UI17 (M2GD_HUMAN)  |
| 280    | MT000199 | Enoyl-CoA hydratase, short chain, 1, mitochondrial              | UniProtKB–P30084 (ECHM_HUMAN)  |
| 281    | MT000945 | Era-like 12S mitochondrial rRNA chaperone 1                     | UniProtKB–O75616 (ERAL1_HUMAN)   |
| 282    | MT000212 | Electron-transfer-flavoprotein, $\alpha$ polypeptide            | UniProtKB–P13804 (ETFA_HUMAN)  |
| 283    | MT000213 | Electron-transfer-flavoprotein, $\beta$ polypeptide             | UniProtKB–P38117 (ETFB_HUMAN), PMID: 8,504,797; PMID: 25,023,281                     |
| 284    | MT000214 | Electron-transferring-flavoprotein dehydrogenase                | UniProtKB–Q16134 (ETFD_HUMAN)  |



**Table 1** (continued)

| Sr. no | MITO ID  | Gene/protein name   | Source   |
|--------|----------|---|--|
| 285    | MT000222 | Ferredoxin 1  | UniProtKB–P10109 (ADX_HUMAN), PMID: 20,547,883   |
| 286    | MT000963 | Glutaminase 2 (liver, mitochondrial)                        | UniProtKB–Q9UI32 (GLSL_HUMAN), PMID: 20,378,837  |
| 287    | MT000255 | Glutamate dehydrogenase 1                                   | UniProtKB–P00367 (DHE3_HUMAN)  |
| 288    | MT000765 | Glycine-N-acyltransferase                                   | UniProtKB–Q6IB77 (GLYAT_HUMAN), PMID: 22,475,485   |
| 289    | MT001421 | Glutamic pyruvate transaminase (alanine aminotransferase) 2 | UniProtKB–Q8TD30 (ALAT2_HUMAN)   |
| 290    | MT000887 | Histidyl-tRNA synthetase 2, mitochondrial                   | UniProtKB–P49590 (SYHM_HUMAN), PMID: 21,464,306  |
| 291    | MT000839 | 3-Hydroxyisobutyrate dehydrogenase                          | UniProtKB–P31937 (3HIDH_HUMAN)   |
| 292    | MT000943 | 3-Hydroxyisobutyryl-CoA hydrolase                           | UniProtKB–Q6NVY1 (HIBCH_HUMAN)   |
| 293    | MT000292 | 3-Hydroxy-3-methylglutaryl-CoA synthase 2 (mitochondrial)   | UniProtKB–P54868 (HMCS2_HUMAN)   |
| 294    | MT000306 | Heat shock 60 kDa protein 1 (chaperonin)                    | UniProtKB–P10809 (CH60_HUMAN)  |
| 295    | MT001171 | Isoleucyl-tRNA synthetase 2, mitochondrial                  | UniProtKB–Q9NSE4 (SYIM_HUMAN)  |
| 296    | MT000309 | Immature colon carcinoma transcript 1                       | UniProtKB–Q14197 (ICT1_HUMAN), PMID: 20,186,120 PMID: 23,908,630                                   |
| 297    | MT000313 | Isocitrate dehydrogenase 3 (NAD <sup>+</sup> ) $\alpha$     | UniProtKB–P50213 (IDH3A_HUMAN)   |
| 298    | MT000314 | Isocitrate dehydrogenase 3 (NAD <sup>+</sup> ) $\beta$      | UniProtKB–O43837 (IDH3B_HUMAN)   |
| 299    | MT000324 | Isovaleryl-CoA dehydrogenase                                | UniProtKB–P26440 (IVD_HUMAN)   |
| 300    | MT000880 | Leucyl-tRNA synthetase 2, mitochondrial                     | UniProtKB–Q15031 (SYLM_HUMAN)  |
| 301    | MT001473 | Methionyl-tRNA synthetase 2, mitochondrial                  | UniProtKB–Q96GW9 (SYMM_HUMAN)  |
| 302    | MT001197 | Methylcrotonoyl-CoA carboxylase 1 ( $\alpha$ )              | UniProtKB–Q96RQ3 (MCCA_HUMAN), PMID: 16,023,992  |
| 303    | MT001254 | Methylcrotonoyl-CoA carboxylase 2 ( $\beta$ )               | UniProtKB–Q9HCC0 (MCCB_HUMAN), PMID: 11,170,888; PMID: 16,023,992                                  |
| 304    | MT001418 | Methylmalonyl CoA epimerase                                 | UniProtKB–Q96PE7 (MCEE_HUMAN)  |
| 305    | MT000350 | Malate dehydrogenase 2, NAD (mitochondrial)                 | UniProtKB–P40926 (MDHM_HUMAN)  |
| 306    | MT001472 | Mitochondrial genome maintenance exonuclease 1              | UniProtKB–Q9BQP7 (MGME1_HUMAN), PMID: 23,313,956 PMID: 23,358,826                                  |
| 307    | MT001575 | Methylmalonic aciduria (cobalamin deficiency) cblA type     | UniProtKB–Q8IVH4 (MMAA_HUMAN)  |
| 308    | MT001634 | Methylmalonic aciduria (cobalamin deficiency) cblB type     | UniProtKB–Q96EY8 (MMAB_HUMAN)  |
| 309    | MT001515 | Mitochondrial ribosomal protein L10                         | UniProtKB–Q7Z7H8 (RM10_HUMAN), PMID: 25,278,503 PMID: 25,838,379 PMID: 28,892,042                  |
| 310    | MT001286 | Mitochondrial ribosomal protein L11                         | UniProtKB–Q9Y3B7 (RM11_HUMAN), PMID: 11,279,069 PMID: 25,278,503 PMID: 25,838,379 PMID: 28,892,042 |
| 311    | MT000541 | Mitochondrial ribosomal protein L12                         | UniProtKB–P52815 (RM12_HUMAN), PMID: 8,626,705   |
| 312    | MT000983 | Mitochondrial ribosomal protein L13                         | UniProtKB–Q9BYD1 (RM13_HUMAN), PMID: 25,278,503 PMID: 25,278,503 PMID: 28,892,042                  |
| 313    | MT000984 | Mitochondrial ribosomal protein L18                         | UniProtKB–Q9H0U6 (RM18_HUMAN), PMID: 25,278,503 PMID: 28,892,042                                   |
| 314    | MT001126 | Mitochondrial ribosomal protein L20                         | UniProtKB–Q9BYC9 (RM20_HUMAN), PMID: 11,279,069 PMID: 25,278,503 PMID: 25,838,379 PMID: 28,892,042 |
| 315    | MT000539 | Mitochondrial ribosomal protein L23                         | UniProtKB–Q16540 (RM23_HUMAN), PMID: 25,278,503 PMID: 25,838,379 PMID: 28,892,042                  |
| 316    | MT001047 | Mitochondrial ribosomal protein L27                         | UniProtKB–Q9P0M9 (RM27_HUMAN), PMID: 11,279,069 PMID: 25,278,503 PMID: 25,838,379 PMID: 28,892,042 |
| 317    | MT000794 | Mitochondrial ribosomal protein L28                         | UniProtKB–Q13084 (RM28_HUMAN), PMID: 25,278,503 PMID: 28,892,042                                   |
| 318    | MT000847 | Mitochondrial ribosomal protein L3                          | UniProtKB–P09001 (RM03_HUMAN), PMID: 25,278,503 PMID: 25,838,379 PMID: 28,892,042                  |
| 319    | MT001285 | Mitochondrial ribosomal protein L32                         | UniProtKB–Q9BYC8 (RM32_HUMAN), PMID: 25,278,503 PMID: 25,838,379 PMID: 28,892,042                  |
| 320    | MT000643 | Mitochondrial transcription termination factor 1            | UniProtKB–Q99551 (MTEF1_HUMAN)   |
| 321    | MT001467 | Mitochondrial ribosome-associated GTPase 1                  | UniProtKB–Q9BT17 (MTG1_HUMAN), PMID: 12,808,030 PMID: 23,396,448                                   |
| 322    | MT000388 | Methylmalonyl CoA mutase                                    | UniProtKB–P22033 (MUTA_HUMAN), PMID: <a href="#">1,970,180</a>                                     |
| 323    | MT001573 | N-acetylglutamate synthase                                  | UniProtKB–Q8N159 (NAGS_HUMAN), PMID: 7,126,172   |

**Table 1** (continued)

| Sr. no | MITO ID  | Gene/protein name   | Source  |
|--------|----------|---|---|
| 324    | MT000403 | NADH dehydrogenase (ubiquinone) 1 $\alpha$ subcomplex, 9, 39 kDa                            | UniProtKB–Q16795 (NDUA9_HUMAN), PMID: 12,611,891                  |
| 325    | MT001652 | NOP2/Sun domain family, member 4  | UniProtKB–Q96CB9 (NSUN4_HUMAN), PMID: 21,531,335 PMID: 22,949,673 |
| 326    | MT000445 | Ornithine aminotransferase  | UniProtKB–P04181 (OAT_HUMAN), PMID: 23,076,989                    |
| 327    | MT000450 | Ornithine carbamoyltransferase  | UniProtKB–P00480 (OTC_HUMAN)                                      |
| 328    | MT000452 | 3-Oxoacid CoA transferase 1   | UniProtKB–P55809 (SCOT1_HUMAN)                                    |
| 329    | MT000458 | Propionyl CoA carboxylase, $\alpha$ polypeptide   | UniProtKB–P05165 (PCCA_HUMAN), PMID: 16,023,992 PMID: 10,101,253  |
| 330    | MT000459 | Propionyl CoA carboxylase, $\beta$ polypeptide  | UniProtKB–P05166 (PCCB_HUMAN), PMID: 16,023,992                   |
| 331    | MT000460 | Phosphoenolpyruvate carboxykinase 2 (mitochondrial)   | UniProtKB–Q16822 (PCKGM_HUMAN)                                    |
| 332    | MT000464 | Pyruvate dehydrogenase (lipoamide) $\beta$  | UniProtKB–P11177 (ODPB_HUMAN)                                     |
| 333    | MT000647 | Pyruvate dehydrogenase complex, component X   | UniProtKB–O00330 (ODPX_HUMAN)                                     |
| 334    | MT000467 | Pyruvate dehydrogenase kinase, isozyme 3  | UniProtKB–Q15120 (PDK3_HUMAN)                                     |
| 335    | MT001225 | Pyruvate dehydrogenase phosphatase catalytic subunit 2                                      | UniProtKB–Q9P2J9 (PDP2_HUMAN)                                     |
| 336    | MT001128 | Pyruvate dehydrogenase phosphatase regulatory subunit                                       | UniProtKB–Q8NCN5 (PDPR_HUMAN)                                     |
| 337    | MT000898 | Prenyl (decaprenyl) diphosphate synthase, subunit 1   | UniProtKB–Q5T2R2 (DPS1_HUMAN)                                     |
| 338    | MT001210 | Prenyl (decaprenyl) diphosphate synthase, subunit 2   | UniProtKB–Q86YH6 (DLP1_HUMAN)                                     |
| 339    | MT000787 | Pitriysin metallopeptidase 1  | UniProtKB–Q5JRX3 (PREP_HUMAN), PMID: 16,849,325 PMID: 19,196,155  |
| 340    | MT000848 | Polymerase (DNA directed), $\gamma$ 2, accessory subunit                                    | UniProtKB–Q9UHN1 (DPOG2_HUMAN)                                    |
| 341    | MT000485 | Polymerase (RNA) mitochondrial (DNA directed)   | UniProtKB–O00411 (RPOM_HUMAN), PMID: 23,283,301 PMID: 29,445,193  |
| 342    | MT000932 | Pentatricopeptide repeat domain 1   | UniProtKB–O75127 (PTCD1_HUMAN), PMID: 19,651,879 PMID: 21,857,155 |
| 343    | MT001208 | Arginyl-tRNA synthetase 2, mitochondrial  | UniProtKB–Q5T160 (SYRM_HUMAN)                                     |
| 344    | MT001104 | Seryl-tRNA synthetase 2, mitochondrial  | UniProtKB–Q9NP81 (SYSM_HUMAN)                                     |
| 345    | MT000034 | Solute carrier family 25 (mitochondrial carrier; adenine nucleotide translocator), member 4 | UniProtKB–P12235 (ADT1_HUMAN)                                     |
| 346    | MT000682 | Succinate-CoA ligase, ADP-forming, $\beta$ subunit  | UniProtKB–Q9P2R7 (SUCB1_HUMAN), PMID: 15,877,282 PMID: 17,287,286 |
| 347    | MT000681 | Succinate-CoA ligase, $\alpha$ subunit  | UniProtKB–P53597 (SUCA_HUMAN)                                     |
| 348    | MT000680 | Succinate-CoA ligase, GDP-forming, $\beta$ subunit  | UniProtKB–Q96199 (SUCB2_HUMAN)                                    |
| 349    | MT001322 | Transcription elongation factor, mitochondrial  | UniProtKB–Q96QE5 (TEFM_HUMAN), PMID: 21,278,163                   |
| 350    | MT000593 | Transcription factor A, mitochondrial   | UniProtKB–Q00059 (TFAM_HUMAN), PMID: 1,737,790                    |
| 351    | MT001258 | Transcription factor B2, mitochondrial  | UniProtKB–Q9H5Q4 (TFB2M_HUMAN)                                    |
| 352    | MT000916 | Thioredoxin 2   | UniProtKB–Q99757 (THIOM_HUMAN), PMID: 12,032,145 PMID: 12,080,052 |
| 353    | MT000796 | Thioredoxin reductase 2   | UniProtKB–Q9NNW7 (TRXR2_HUMAN), PMID: 10,215,850                  |
| 354    | MT000772 | Tryptophanyl tRNA synthetase 2, mitochondrial   | UniProtKB–Q9UGM6 (SYWM_HUMAN), PMID: 28,236,339                   |
| 355    | MT001021 | Tyrosyl-tRNA synthetase 2, mitochondrial  | UniProtKB–Q9Y2Z4 (SYYM_HUMAN), PMID: 15,779,907                   |
| 356    | MT000051 | ATP synthase, H <sup>+</sup> transporting, mitochondrial F1 complex, $\beta$ polypeptide    | UniProtKB–P06576 (ATPB_HUMAN)                                     |
| 357    | MT000052 | ATP synthase, H <sup>+</sup> transporting, mitochondrial F1 complex, $\gamma$ polypeptide 1 | UniProtKB–P36542 (ATPG_HUMAN)                                     |
| 358    | MT000802 | Phenylalanyl-tRNA synthetase 2, mitochondrial   | UniProtKB–O95363 (SYFM_HUMAN)                                     |
| 359    | MT000940 | Mitochondrial ribosome-associated GTPase 2  | UniProtKB–Q9H4K7 (MTG2_HUMAN)                                     |
| 360    | MT000882 | Sirtuin 4   | UniProtKB–Q9Y6E7 (SIR4_HUMAN)                                     |

**Table 2** List of proteins that are not present exclusively in mitochondria

| Sr. no | Mitoproteome ID | Gene/protein name   | Location  | Source  |
|--------|-----------------|---|---|---|
| 1      | MT000745        | ATP-binding cassette, subfamily B (MDR/TAP), member 6 (Langereis blood group) | Golgi apparatus/ER/PM/OM                        | UniProtKB–Q9NP58 (ABCB6_HUMAN)                                      |
| 2      | MT000216        | Acyl-CoA synthetase long-chain family member 1                                | OM/Peroxisome/ER                                | UniProtKB–P33121 (ACSL1_HUMAN)                                      |
| 3      | MT000217        | Acyl-CoA synthetase long-chain family member 3                                | OM/Peroxisome/ER                                | Gene ID: 2181 & UniProtKB–O95573 (ACSL3_HUMAN)                      |
| 4      | MT000218        | Acyl-CoA synthetase long-chain family member 4                                | OM/Peroxisome/ER/PM                             | Gene ID: 2182 & UniProtKB–O60488 (ACSL4_HUMAN)                      |
| 5      | MT001070        | Acyl-CoA synthetase long-chain family member 5                                | PM/ER/OM  | Gene ID: 51,703 & UniProtKB–Q9ULC5 (ACSL5_HUMAN)                    |
| 6      | MT000879        | Acyl-CoA synthetase long-chain family member 6                                | Peroxisome/ER/OM                                | Gene ID: 51,703<br>UniProtKB–Q9ULC5 (ACSL5_HUMAN)                   |
| 7      | MT001179        | ArfGAP with dual PH domains 2   | Cytoplasm and PM                                | Gene ID: 55,803<br>Human protein ATLAS (ADAP2)                      |
| 8      | MT001154        | 1-Acylglycerol-3-phosphate O-acyl-transferase 5                               | ER/Nucleus/OM                                   | UniProtKB–Q9NUQ2 (PLCE_HUMAN)                                       |
| 9      | MT001430        | Apoptosis-inducing factor, mitochondrion-associated, 2                        | Cytoplasm/PM/Nucleus/OM                         | UniProtKB–Q9BRQ8 (FSP1_HUMAN)                                       |
| 10     | MT000021        | V-akt murine thymoma viral oncogene homolog 1                                 | PM/Nucleus/Cytoplasm                            | UniProtKB–P31749 (AKT1_HUMAN)                                       |
| 11     | MT001061        | Armadillo repeat containing, X-linked 3                                       | Cytoplasm/OM/Nucleus                            | UniProtKB–Q9UH62 (ARMX3_HUMAN)                                      |
| 12     | MT000045        | Argininosuccinate synthase 1  | Cytosol   | UniProtKB–P00966 (ASSY_HUMAN)<br>Human protein ATLAS (ADAP2)        |
| 13     | MT000150        | Activating transcription factor 2   | OM/Nucleus/Cytoplasm                            | UniProtKB–P15336 (ATF2_HUMAN)                                       |
| 14     | T000068         | BCL2-associated agonist of cell death   | OM/Cytoplasm                                    | UniProtKB–Q92934 (BAD_HUMAN)  |
| 15     | MT000076        | B-cell CLL/lymphoma 2   | ER/Nucleus/OM                                   | UniProtKB–P10415 (BCL2_HUMAN)                                       |
| 16     | MT000077        | BCL2-related protein A1   | Cytoplasm                                       | UniProtKB–Q16548 (B2LA1_HUMAN)                                      |
| 17     | MT000078        | BCL2-like 1   | OM/Cytoplasm/Nucleus/Centrosome                 | UniProtKB–Q07817 (B2CL1_HUMAN)                                      |
| 18     | MT000743        | BCL2-like 10 (apoptosis facilitator)  | ER/OM/Nucleus                                   | UniProtKB–Q9HD36 (B2L10_HUMAN)                                      |
| 19     | MT000744        | BCL2-like 11 (apoptosis facilitator)  | Endomembrane system/OM/IMS                      | UniProtKB–O43521 (B2L11_HUMAN)                                      |
| 20     | MT000079        | BCL2-like 2   | Mitochondrion/Peripheral membrane               | UniProtKB–Q92843 (B2CL2_HUMAN)                                      |
| 21     | MT000084        | BH3-interacting domain death agonist  | Cytoplasm/OM                                    | UniProtKB–P55957 (BID_HUMAN)  |
| 22     | MT000085        | BCL2-interacting killer (apoptosis-inducing)                                  | Endomembrane system/OM/IMS                      | UniProtKB–Q13323 (BIK_HUMAN)  |
| 23     | MT001451        | Bcl2 modifying factor   | Cytoplasm/OM/PM/Cytoskeleton/Myosin complex     | UniProtKB–Q96LC9 (BMF_HUMAN)  |
| 24     | MT000088        | BCL2/adenovirus E1B 19 kDa-interacting protein 3-like                         | OM/ER/Nucleus                                   | UniProtKB–O60238 (BNI3L_HUMAN)                                      |
| 25     | MT000089        | BCL2-related ovarian killer   | GolgiApparatus/ER/Nucleus/OM/Cytoplasm/Endosome | UniProtKB–Q9UMX3 (BOK_HUMAN)  |
| 26     | MT000100        | Calnexin  | ER/Melanosome                                   | Gene ID: 821 & UniProtKB–P27824 (CALX_HUMAN)<br>Human protein ATLAS |
| 27     | MT000106        | Caspase 8, apoptosis-related cysteine peptidase                               | Cytoplasm and Nucleus                           | UniProtKB–Q14790 (CASP8_HUMAN)<br>Human protein ATLAS               |
| 28     | MT000118        | Cell death-inducing DFFA-like effector a                                      | Nucleus/Lipid droplet                           | UniProtKB–O60543 (CIDEA_HUMAN)                                      |
| 29     | MT001678        | CDGSH iron sulfur domain 2  | ER/OM   | Gene ID: 493,856<br>Human protein ATLAS                             |
| 30     | MT000637        | Zinc finger protein 205   | Nucleus   | UniProtKB–O95201 (ZN205_HUMAN)<br>Human protein ATLAS               |
| 31     | MT001357        | CoA synthase  | MM/Cytoplasm                                    | UniProtKB–Q13057 (COASY_HUMAN)                                      |
| 32     | MT001521        | Carnitine palmitoyltransferase 1C   | OM and ER                                       | UniProtKB–Q8TCG5 (CPT1C_HUMAN)                                      |

**Table 2** (continued)

| Sr. no | Mitoproteome ID | Gene/protein name  | Location                                    | Source   |
|--------|-----------------|--|---|--|
| 33     | MT000159        | Cytochrome b5 type A (microsomal)  | ER/cytoplasm                                | UniProtKB-P00167 (CYB5_HUMAN)<br>Human protein ATLAS             |
| 34     | MT000183        | Cytochrome b5 reductase 3  | ER/OM/Cytoplasm                             | Gene ID: 1727  |
| 35     | MT001077        | Zinc finger, FYVE domain containing 1  | Mitochondrion/ER/Golgi Apparatus            | UniProtKB-Q9HBF4 (ZFYV1_HUMAN)                                   |
| 36     | MT000178        | DEAD (Asp-Glu-Ala-Asp) box helicase 3, X-linked  | PM/Nucleus/Cytoplasm/Centrosome             | UniProtKB-O00571 (DDX3X_HUMAN)<br>Human protein ATLAS            |
| 37     | MT000189        | Dystrophia myotonica-protein kinase  | Cytoplasm/Nucleus/ER/PM/OM                  | UniProtKB-Q09013 (DMPK_HUMAN)                                    |
| 38     | MT000746        | Dynamin 1-like   | Peroxisome/Golgi Apparatus/OM and cytoplasm | UniProtKB-O00429 (DNM1L_HUMAN)                                   |
| 39     | MT000667        | Dynein, light chain, LC8-type 1  | Mitochondrion/Cytoskeleton and Nucleus      | UniProtKB-P63167 (DYL1_HUMAN)                                    |
| 40     | MT001552        | Dynein, light chain, LC8-type 2  | Cytoskeleton                                | UniProtKB-Q96FJ2 (DYL2_HUMAN)<br>Human protein ATLAS             |
| 41     | MT000196        | E2F transcription factor 1   | Nucleus                                     | UniProtKB-Q01094 (E2F1_HUMAN)<br>Human protein ATLAS             |
| 42     | MT000207        | EPH receptor A4  | PM/early endosome                           | UniProtKB-P54764 (EPHA4_HUMAN)                                   |
| 43     | MT001016        | Fission 1 (mitochondrial outer membrane) homolog ( <i>S. cerevisiae</i> )  | Peroxisome/OM                               | UniProtKB-Q9Y3D6 (FIS1_HUMAN)                                    |
| 44     | MT001082        | Ganglioside-induced differentiation-associated protein 1   | OM and Cytoplasm                            | UniProtKB-Q8TB36 (GDAP1_HUMAN)                                   |
| 45     | MT001156        | GTPase, IMAP family member 5   | Lysosome and Endosome                       | UniProtKB-Q96F15 (GIMA5_HUMAN)                                   |
| 46     | MT000246        | Gap junction protein, $\alpha$ 1, 43 kDa   | ER and PM                                   | UniProtKB-P17302 (CXA1_HUMAN)                                    |
| 47     | MT000247        | Glycerol kinase  | OM and Cytoplasm                            | UniProtKB-P32189 (GLPK_HUMAN)                                    |
| 48     | MT000248        | Glycerol kinase 2  | OM and Cytoplasm                            | UniProtKB-Q14410 (GLPK2_HUMAN)                                   |
| 49     | MT000269        | Glycogen synthase kinase 3 $\alpha$  | Cytosol/Cytoskeleton/OM/Nucleus             | UniProtKB-P49840 (GSK3A_HUMAN)                                   |
| 50     | MT000270        | Glycogen synthase kinase 3 $\beta$   | PM/Nucleus/cytoplasm                        | UniProtKB-P49841 (GSK3B_HUMAN)                                   |
| 51     | MT001360        | Cytochrome b5 type B (outer mitochondrial membrane)  | OM/Other species Pan troglodytes            | Gene ID: 80,777 & UniProtKB-O43169 (CYB5B_HUMAN)<br>NCBI Blast P |
| 52     | MT000274        | Granzyme B (granzyme 2, cytotoxic T-lymphocyte-associated serine esterase 1)                                       | Lysosome/Secreted Extracellular region      | UniProtKB-P10144 (GRAB_HUMAN)                                    |
| 53     | MT000278        | Hydroxyacyl-CoA dehydrogenase/3-ketoacyl-CoA thiolase/enoyl-CoA hydratase (trifunctional protein), $\beta$ subunit | OM and ER                                   | UniProtKB-P55084 (ECHB_HUMAN)                                    |
| 54     | MT000287        | Hexokinase 1   | Cytosol and OM                              | UniProtKB-P19367 (HXK1_HUMAN)                                    |
| 55     | MT000288        | Hexokinase 2   | Cytosol and OM                              | UniProtKB-P52789 (HXK2_HUMAN)                                    |
| 56     | MT000505        | Mitogen-activated protein kinase 8   | Nucleus and cytoplasm                       | UniProtKB-P45983 (MK08_HUMAN)                                    |
| 57     | MT001266        | Mitochondrial amidoxime-reducing component 1   | OM and Other species Pan paniscus           | UniProtKB-Q5VT66 (MARC1_HUMAN)<br>NCBI Blast P                   |
| 58     | MT001116        | Mitochondrial amidoxime-reducing component 2   | OM and Peroxisome                           | UniProtKB-Q969Z3 (MARC2_HUMAN)                                   |
| 59     | MT001096        | Membrane-associated ring finger (C3HC4) 5  | OM and ER                                   | UniProtKB-Q9NX47 (MARH5_HUMAN)                                   |
| 60     | MT001223        | Mitochondrial antiviral signaling protein  | OM and Peroxisome                           | UniProtKB-Q7Z434 (MAVS_HUMAN)                                    |
| 61     | MT000347        | Myeloid cell leukemia 1  | OM/Cytoplasm and Nucleoplasm                | UniProtKB-Q07820 (MCL1_HUMAN)                                    |
| 62     | MT001200        | Mitochondrial fission factor   | OM/Peroxisome/Synaptic vesicle              | UniProtKB-Q9GZY8 (MFF_HUMAN)                                     |
| 63     | MT001168        | Mitofusin 1  | OM and Cytoplasm                            | UniProtKB-Q8IWA4 (MFN1_HUMAN)                                    |
| 64     | MT000859        | MLX-interacting protein  | OM/Nucleus/Cytoplasm                        | UniProtKB-Q9HAP2 (MLXIP_HUMAN)                                   |
| 65     | MT000357        | Microsomal glutathione S-transferase 1   | OM and ER                                   | UniProtKB-P10620 (MGST1_HUMAN)                                   |
| 66     | MT001255        | Modulator of apoptosis 1   | OM, Cytoplasm, Cytosol                      | UniProtKB-Q96BY2 (MOAP1_HUMAN)                                   |
| 67     | MT001610        | MPV17 mitochondrial membrane protein-like  | Peroxisome membrane                         | UniProtKB-Q2QL34 (MP17L_HUMAN)                                   |

**Table 2** (continued)

| Sr. no | Mitoproteome ID | Gene/protein name   | Location   | Source  |
|--------|-----------------|---|--|---|
| 68     | MT001136        | Misato 1, mitochondrial distribution and morphology regulator   | OM and Cytoplasm                                 | UniProtKB-Q9BUK6 (MSTO1_HUMAN)                  |
| 69     | MT000233        | Mechanistic target of rapamycin (serine/threonine kinase)       | OM/ER/Golgi Apparatus/Nucleus/Cytoplasm          | UniProtKB-P42345 (MTOR_HUMAN)                   |
| 70     | MT000387        | Metaxin 1   | OM/Other Membrane                                | UniProtKB-Q13505 (MTX1_HUMAN)                   |
| 71     | MT001313        | Mitochondrial E3 ubiquitin protein ligase 1                     | OM and Peroxisome                                | UniProtKB-Q969V5 (MUL1_HUMAN)                   |
| 72     | MT001344        | Myosin XIX  | OM and Cytoskeleton                              | UniProtKB-Q96H55 (MYO19_HUMAN)                  |
| 73     | MT000392        | Myocilin, trabecular meshwork inducible glucocorticoid response | ER, OM, IM, Golgi Apparatus/Cytoplasmic vesicles | UniProtKB-Q99972 (MYOC_HUMAN)                   |
| 74     | MT000435        | N-myristoyltransferase 1  | Cytoplasm and other membrane                     | UniProtKB-P30419 (NMT1_HUMAN)                   |
| 75     | MT000472        | Progesterone receptor   | OM/Nucleus/Cytoplasm                             | UniProtKB-P06401 (PRGR_HUMAN)                   |
| 76     | MT000476        | Phosphatidylinositol 4-kinase, catalytic, $\beta$               | OM/ER/Golgi Apparatus                            | UniProtKB-Q9UBF8 (PI4KB_HUMAN)                  |
| 77     | MT001289        | PTEN-induced putative kinase 1                                  | OM and Cytosol                                   | UniProtKB-Q9BXM7 (PINK1_HUMAN)                  |
| 78     | MT000495        | Protein phosphatase 2, regulatory subunit B, $\beta$            | OM and Cytoplasm, cytoskeleton                   | UniProtKB-Q00005 (2ABB_HUMAN)                   |
| 79     | MT000497        | Protein phosphatase 3, catalytic subunit, $\gamma$ isozyme      | Cytosol and Mitochondrion                        | UniProtKB-P48454 (PP2BC_HUMAN)                  |
| 80     | MT000498        | Protein phosphatase 3, regulatory subunit B, $\alpha$           | PM and Cytosol                                   | UniProtKB-P63098 (CANB1_HUMAN)                  |
| 81     | MT000934        | RAB11 family-interacting protein 5 (class I)                    | Golgi Apparatus, OM, cytoplasm and endosome      | UniProtKB-Q9BXF6 (RFIP5_HUMAN)                  |
| 82     | MT000532        | Raf-1 proto-oncogene, serine/threonine kinase                   | PM/Nucleus/Cytoplasm/OM                          | UniProtKB-P04049 (RAF1_HUMAN)                   |
| 83     | MT001141        | Regulator of microtubule dynamics 3                             | OM/Nucleus/Cytoplasm/Cytoskeleton                | UniProtKB-Q96TC7 (RMD3_HUMAN)                   |
| 84     | MT001459        | Ring finger protein 185   | ER and OM  | UniProtKB-Q96GF1 (RN185_HUMAN)                  |
| 85     | MT000544        | Ribosomal protein S6 kinase, 70 kDa, polypeptide 1              | OM/Nucleus/Cytoplasm                             | UniProtKB-P23443 (KS6B1_HUMAN)                  |
| 86     | MT001460        | Radical S-adenosyl methionine domain containing 2               | OM/ER/Golgi Apparatus                            | UniProtKB-Q8WXG1 (RSAD2_HUMAN)                  |
| 87     | MT000913        | SAMM50 sorting and assembly machinery component                 | OM/Cytoplasm                                     | UniProtKB-Q9Y512 (SAM50_HUMAN)                  |
| 88     | MT000260        | Stratifin   | Nucleus and cytoplasm                            | UniProtKB-P31947 (1433S_HUMAN)                  |
| 89     | MT001028        | SH3-domain GRB2-like endophilin B1                              | OM/Cytoplasm/Golgi Apparatus                     | UniProtKB-Q9Y371 (SHLB1_HUMAN)                  |
| 90     | MT000793        | SIVA1, apoptosis-inducing factor                                | Nucleus and cytoplasm                            | UniProtKB-O15304 (SIVA_HUMAN)                   |
| 91     | MT001112        | Solute carrier family 35, member F6                             | Lysosome and OM                                  | UniProtKB-Q8N357 (S35F6_HUMAN)                  |
| 92     | MT000888        | Solute carrier family 44 (choline transporter), member 1        | PM and OM  | UniProtKB-Q8WWI5 (CTL1_HUMAN)                   |
| 93     | MT000573        | SRY (sex determining region Y)-box 10                           | OM, Nucleus and cytoplasm                        | UniProtKB-P56693 (SOX10_HUMAN)                  |
| 94     | MT001537        | Spermatogenesis-associated 18                                   | OM and cytoplasm                                 | UniProtKB-Q8TC71 (MIEAP_HUMAN)                  |
| 95     | MT000868        | Spastic paraplegia 20 (Troyer syndrome)                         | Cytosol and Midbody                              | UniProtKB-A0A024RDV9 (A0A024RDV9_HUMAN)         |
| 96     | MT001122        | Syntaxin 17   | ER, Cytosol                                      | UniProtKB-P56962 (STX17_HUMAN)                  |
| 97     | MT000594        | Transcription factor Dp-1                                       | Nucleus and cytoplasm                            | UniProtKB-Q14186 (TFDP1_HUMAN)                  |
| 98     | MT001623        | Transmembrane protein 102                                       | Cell Membrane                                    | UniProtKB-Q8N9M5 (TM102_HUMAN)                  |
| 99     | MT001638        | Transmembrane protein 173                                       | OM, PM and ER                                    | UniProtKB-Q86WV6 (STING_HUMAN)                  |
| 100    | MT000821        | Translocase of outer mitochondrial membrane 34                  | OM and Cytoplasm                                 | Gene ID: 10,953, UniProtKB-Q15785 (TOM34_HUMAN) |
| 101    | MT000600        | Tumor protein p53   | ER, MM, Cytoplasm, Centrosome, Nucleus           | UniProtKB-P04637 (P53_HUMAN)                    |
| 102    | MT000094        | Translocator protein (18 kDa)                                   | MM, PM   | UniProtKB-P30536 (TSPO_HUMAN)                   |
| 103    | MT000588        | Vesicle-associated membrane protein 1 (synaptobrevin 1)         | OM, Cytoplasmic/Synaptic vesicle                 | UniProtKB-P23763 (VAMP1_HUMAN)                  |

**Table 2** (continued)

| Sr. no | Mitoproteome ID | Gene/protein name   | Location  | Source  |
|--------|-----------------|---|---|---|
| 104    | MT000786        | Vesicle amine transport 1   | OM, Cytoplasm   | UniProtKB-Q99536 (VAT1_HUMAN)                   |
| 105    | MT000625        | Voltage-dependent anion channel 1   | PM, OM  | UniProtKB-P21796 (VDAC1_HUMAN)                  |
| 106    | MT000631        | Tyrosine 3-monooxygenase/tryptophan 5-monooxygenase activation protein, $\beta$             | Cytoplasm, Melanosome   | UniProtKB-P31946 (1433B_HUMAN)                  |
| 107    | MT000632        | Tyrosine 3-monooxygenase/tryptophan 5-monooxygenase activation protein, $\epsilon$          | Nucleus, cytoplasm, Melanosome                                | UniProtKB-P62258 (1433E_HUMAN)                  |
| 108    | MT000633        | Tyrosine 3-monooxygenase/tryptophan 5-monooxygenase activation protein, $\gamma$            | Cytoplasm   | UniProtKB-P61981 (1433G_HUMAN)                  |
| 109    | MT000634        | Tyrosine 3-monooxygenase/tryptophan 5-monooxygenase activation protein, $\eta$              | Cytosol, Mitochondrion, Extracellular exosome, PM, Cytoplasm  | UniProtKB-Q04917 (1433F_HUMAN)                  |
| 110    | MT000824        | Tyrosine 3-monooxygenase/tryptophan 5-monooxygenase activation protein, $\theta$            | Cytoplasm   | UniProtKB-P27348 (1433T_HUMAN)                  |
| 111    | MT000635        | Tyrosine 3-monooxygenase/tryptophan 5-monooxygenase activation protein, $\zeta$             | Cytoplasm, Melanosome   | UniProtKB-P63104 (1433Z_HUMAN)                  |
| 112    | MT001181        | CDGSH iron sulfur domain 1  | OM, Other species pan troglodytes                             | UniProtKB-Q9NZ45 (CISD1_HUMAN)<br>NCBI Blast P  |
| 113    | MT001476        | ATPase inhibitory factor 1  | OM, Other species Pongo abelii                                | UniProtKB-Q9UII2 (ATIF1_HUMAN)<br>NCBI Blast P  |
| 114    | MT001084        | Mitochondrial elongation factor 1   | OM, Other species Xenopus tropicalis                          | UniProtKB-Q9NQG6 (MID51_HUMAN)<br>NCBI Blast P  |
| 115    | MT000035        | Solute carrier family 25 (mitochondrial carrier; adenine nucleotide translocator), member 5 | IM, PM  | UniProtKB-P05141 (ADT2_HUMAN)                   |
| 116    | MT000651        | Stannin   | OM, Other species Macaca mulatta                              | UniProtKB-O75324 (SNN_HUMAN)<br>NCBI Blast P    |
| 117    | MT001155        | Synaptojanin 2-binding protein  | OM, other species Pan troglodytes                             | UniProtKB-P57105 (SYJ2B_HUMAN)<br>NCBI Blast P  |
| 118    | MT000734        | Translocase of outer mitochondrial membrane 20 homolog (yeast)                              | OM, Other species Bos mutus                                   | UniProtKB-Q15388 (TOM20_HUMAN)<br>NCBI Blast P  |
| 119    | MT000627        | Voltage-dependent anion channel 3   | OM, PM  | UniProtKB-Q9Y277 (VDAC3_HUMAN)                  |
| 120    | MT000317        | Interferon, $\alpha$ -inducible protein 27  | Mitochondrion, Nucleus, ER                                    | UniProtKB-P40305 (IFI27_HUMAN)                  |
| 121    | MT000448        | Optic atrophy 1 (autosomal dominant)  | Present between ER and mitochondrion membrane                 | UniProtKB-O60313 (OPA1_HUMAN)                   |
| 122    | MT000686        | Immediate early response 3  | PM  | UniProtKB-P46695 (IEX1_HUMAN)                   |
| 123    | MT000747        | ATP-binding cassette, subfamily F (GCN20), member 2   | PM, Other species Pan troglodytes                             | UniProtKB-Q9UG63 (ABCF2_HUMAN)<br>NCBI Blast P  |
| 124    | MT001090        | Translocase of outer mitochondrial membrane 7 homolog (yeast)                               | PM  | UniProtKB-Q75MR5 (Q75MR5_HUMAN)                 |
| 125    | MT001203        | Translocase of outer mitochondrial membrane 22 homolog (yeast)                              | OM, Other species Macaca mulatta                              | UniProtKB-Q549C5 (Q549C5_HUMAN)<br>NCBI Blast P |
| 126    | MT001667        | Translocase of outer mitochondrial membrane 5 homolog (yeast)                               | OM, Other species Neomonachus schauinslandi                   | UniProtKB-Q8N4H5 (TOM5_HUMAN)<br>NCBI Blast P   |
| 127    | MT001695        | Translocase of outer mitochondrial membrane 6 homolog (yeast)                               | OM, Other species Pan paniscus                                | UniProtKB-Q96B49 (TOM6_HUMAN)<br>NCBI Blast P   |
| 128    | MT000696        | Apoptosis-inducing factor, mitochondrion-associated, 1                                      | IMS, Nucleus, Cytoplasm, IM, MIM, Cytosol, Perinuclear region | UniProtKB-O95831 (AIFM1_HUMAN)                  |
| 129    | MT000042        | ADP-ribosylation factor-like 2  | IMS, Nucleus, Cytoplasm, Centrosome                           | UniProtKB-P36404 (ARL2_HUMAN)                   |
| 130    | MT000896        | ADP-ribosylation factor-like 2-binding protein  | IMS, Nucleus, Cytoplasm, Centrosome                           | UniProtKB-Q9Y2Y0 (AR2BP_HUMAN)                  |
| 131    | MT000043        | Arrestin, $\beta$ 2   | PM, Nucleus, Cytoplasm  | UniProtKB-P32121 (ARRB2_HUMAN)                  |
| 132    | MT000797        | Thioredoxin-interacting protein   | Cytoplasm   | UniProtKB-Q9H3M7 (TXNIP_HUMAN)                  |

**Table 2** (continued)

| Sr. no | Mitoproteome ID | Gene/protein name   | Location   | Source   |
|--------|-----------------|---|--|--|
| 133    | MT000243        | Biogenesis of lysosomal organelles complex-1, subunit 1   | Lysosome, Mitochondrion, Cytosol                   | UniProtKB-P78537 (BL1S1_HUMAN)                                   |
| 134    | MT000109        | Catalase  | Peroxisome   | UniProtKB-P04040 (CATA_HUMAN)                                    |
| 135    | MT001207        | Cytokine-induced apoptosis inhibitor 1  | Nucleus, cytoplasm, IMS                            | UniProtKB-Q6F181 (CPIN1_HUMAN)                                   |
| 136    | MT000748        | COX17 cytochrome c oxidase copper chaperone   | IMS and cytoplasm                                  | UniProtKB-Q14061 (COX17_HUMAN)                                   |
| 137    | MT000194        | Deoxythymidylate kinase (thymidylate kinase)  | Cytosol, IMS, Nucleus, MM                          | UniProtKB-P23919 (KTHY_HUMAN)                                    |
| 138    | MT000942        | F-box and leucine-rich repeat protein 4   | IMS, Nucleus, Cytoplasm                            | UniProtKB-Q9UKA2 (FBXL4_HUMAN)                                   |
| 139    | MT000232        | Frataxin  | Cytosol, Mitochondrion                             | UniProtKB-Q16595 (FRDA_HUMAN)                                    |
| 140    | MT000238        | Glycine amidinotransferase (L-arginine/glycine amidinotransferase)                              | IMS, MIM, MM and cytoplasm                         | UniProtKB-P50440 (GATM_HUMAN)                                    |
| 141    | MT000264        | Glutathione peroxidase 1  | Cytoplasm  | UniProtKB-P07203 (GPX1_HUMAN)                                    |
| 142    | MT000284        | Hepatocyte growth factor (hepatoinetin A; scatter factor)                                       | Extracellular regions, PM                          | UniProtKB-P08581 (MET_HUMAN)                                     |
| 143    | MT000674        | Harakiri, BCL2-interacting protein  | Mitochondrion, PM                                  | UniProtKB-O00198 (HRK_HUMAN)                                     |
| 144    | MT000294        | Hydroxy- $\delta$ -5-steroid dehydrogenase, 3 $\beta$ - and steroid $\delta$ -isomerase 1       | ER, Mitochondrion                                  | UniProtKB-P14060 (3BHS1_HUMAN)                                   |
| 145    | MT000295        | Hydroxy- $\delta$ -5-steroid dehydrogenase, 3 $\beta$ - and steroid $\delta$ -isomerase 2       | ER, Mitochondrial membrane                         | UniProtKB-P26439 (3BHS2_HUMAN)                                   |
| 146    | MT000338        | lamin A/C   | Nucleus  | UniProtKB-P02545 (LMNA_HUMAN)                                    |
| 147    | MT000340        | LYN proto-oncogene, Src family tyrosine kinase  | PM, Golgi Apparatus, Nucleus, Cytoplasm            | UniProtKB-P07948 (LYN_HUMAN)                                     |
| 148    | MT000881        | Sirtuin 5   | IMS, MIM, Nucleus, Cytoplasm                       | UniProtKB-Q9NXA8 (SIR5_HUMAN)                                    |
| 149    | MT000822        | Myeloid/lymphoid or mixed-lineage leukemia (trithorax homolog, Drosophila); translocated to, 11 | Nucleus, cytoplasm, Centrosome                     | UniProtKB-Q13015 (AF1Q_HUMAN)                                    |
| 150    | MT000456        | Parkin RBR E3 ubiquitin protein ligase  | ER, OM, Nucleus, Cytosol                           | UniProtKB-O60260 (PRKN_HUMAN)                                    |
| 151    | MT001162        | Presenilin-associated, rhomboid-like  | MIM, Nucleus                                       | UniProtKB-Q9H300 (PARL_HUMAN)                                    |
| 152    | MT000653        | Phospholipase A2, group VI (cytosolic, calcium-independent)                                     | PM, Mitochondrion, Cytoplasm                       | UniProtKB-O60733 (PLPL9_HUMAN)                                   |
| 153    | MT001445        | Polyribonucleotide nucleotidyltransferase 1   | MIM Space, cytoplasm, MM                           | UniProtKB-Q8TCS8 (PNPT1_HUMAN)                                   |
| 154    | MT001205        | ACN9 homolog ( <i>S. cerevisiae</i> ) MM  | OM, Other species <i>Saccharomyces pastorianus</i> | UniProtKB-Q9NRP4 (SDHF3_HUMAN)<br>NCBI Blast P                   |
| 155    | MT000517        | Proteasome (prosome, macropain) 26S subunit, non-ATPase, 10                                     | Nucleus, cytoplasm                                 | UniProtKB-O75832 (PSD10_HUMAN)                                   |
| 156    | MT000991        | PYD and CARD domain containing  | ER, Mitochondrion, Nucleus, Cytoplasm              | UniProtKB-Q9ULZ3 (ASC_HUMAN)                                     |
| 157    | MT000558        | Serine hydroxymethyltransferase 2 (mitochondrial)   | IM, Nucleus, Cytoplasm                             | UniProtKB-P34897 (GLYM_HUMAN)                                    |
| 158    | MT001057        | TP53 regulated inhibitor of apoptosis 1   | Perinuclear region, IMS                            | UniProtKB-O43715 (TRIA1_HUMAN)                                   |
| 159    | MT000569        | Superoxide dismutase 1, soluble   | Mitochondrion, Nucleus, Cytoplasm                  | UniProtKB-P00441 (SODC_HUMAN)                                    |
| 160    | MT001001        | Stomatin (EPB72)-like 2   | PM, Cytoskeleton, MIM                              | UniProtKB-Q9UJZ1 (STML2_HUMAN)                                   |
| 161    | MT000019        | Adenylate kinase 2  | IMS, other species <i>Pongo abelii</i>             | Gene ID: 204 & UniProtKB-P54819 (KAD2_HUMAN)<br>NCBI Blast P     |
| 162    | MT001081        | Cytochrome c, somatic   | MIM, other species <i>Pan troglodytes</i>          | Gene ID: 54,205 & UniProtKB-P99999 (CYC_HUMAN),<br>NCBI Blast P  |
| 163    | MT000929        | HIG1 hypoxia inducible domain family, member 1A   | MIM, other species <i>Pan troglodytes</i>          | Gene ID: 25,994 & UniProtKB-Q9Y241 (HIG1A_HUMAN)<br>NCBI Blast P |
| 164    | MT000402        | NADH dehydrogenase (ubiquinone) 1 $\alpha$ subcomplex, 8, 19 kDa                                | MIM, other species <i>Pan troglodytes</i>          | Gene ID: 4702 & UniProtKB-P51970 (NDUA8_HUMAN),<br>NCBI Blast P  |

**Table 2** (continued)

| Sr. no | Mitoproteome ID | Gene/protein name   | Location  | Source  |
|--------|-----------------|---|---|---|
| 165    | MT000490        | Protoporphyrinogen oxidase (PPOX)   | MIM, other species Pan troglodytes                | Gene ID: 5498 & UniProtKB–P50336 (PPOX_HUMAN)<br>NCBI Blast P |
| 166    | MT000675        | Tumor necrosis factor (ligand) superfamily, member 10   | PM, Extracellular region                          | UniProtKB–P50591 (TNF10_HUMAN)                                |
| 167    | MT000939        | ATP-binding cassette, subfamily A (ABC1), member 12   | Golgi apparatus, Secretory vesicle                | UniProtKB–Q86UK0 (ABCAC_HUMAN)                                |
| 168    | MT000771        | ATP-binding cassette, subfamily A (ABC1), member 8  | PM  | UniProtKB–O94911 (ABCA8_HUMAN)                                |
| 169    | MT000522        | ATP-binding cassette, subfamily D (ALD), member 3   | Peroxisome membrane                               | UniProtKB–P28288 (ABCD3_HUMAN)                                |
| 170    | MT000029        | Aldehyde dehydrogenase 3 family, member A2  | ER  | UniProtKB–P51648 (AL3A2_HUMAN)                                |
| 171    | MT000969        | APEX nuclease (apurinic/aprimidinic endonuclease) 2   | Mitochondrion, Nucleus, Cytoplasm                 | UniProtKB–Q9UBZ4 (APEX2_HUMAN)                                |
| 172    | MT000049        | ATP synthase, H <sup>+</sup> transporting, mitochondrial F1 complex, $\alpha$ subunit 1, cardiac muscle | MIM, PM   | UniProtKB–P25705 (ATPA_HUMAN)                                 |
| 173    | MT000082        | Brain-derived neurotrophic factor   | Extracellular region secreted                     | UniProtKB–P23560 (BDNF_HUMAN)                                 |
| 174    | MT001578        | Basic helix-loop-helix family, member $\alpha$ 15   | Nucleus   | UniProtKB–Q7RTS1 (BHA15_HUMAN)                                |
| 175    | MT001442        | Calcium-binding protein, spermatid-specific 1   | MIM, Cytoplasm, Flagellum                         | UniProtKB–Q96KC9 (CABS1_HUMAN)                                |
| 176    | MT000676        | CDP-diacylglycerol synthase (phosphatidate cytidyltransferase) 2  | ER  | UniProtKB–O95674 (CDS2_HUMAN)                                 |
| 177    | MT001101        | Coiled-coil-helix-coiled-coil-helix domain containing 3   | MIM, Nucleus, Cytoplasm                           | UniProtKB–Q9NX63 (MIC19_HUMAN)                                |
| 178    | MT000123        | Chloride intracellular channel 1  | PM, Nucleus, Cytoplasm                            | UniProtKB–O00299 (CLIC1_HUMAN)                                |
| 179    | MT001583        | IMP1 inner mitochondrial membrane peptidase-like ( <i>S. cerevisiae</i> )                               | MIM, Other species Pan troglodytes                | UniProtKB–Q96LU5 (IMP1L_HUMAN)<br>NCBI Blast P                |
| 180    | MT000145        | Carbamoyl-phosphate synthase 1, mitochondrial   | Mitochondrion, Nucleolus                          | UniProtKB–P31327 (CPSM_HUMAN)<br>The Human protein ATLAS      |
| 181    | MT000148        | Carnitine palmitoyltransferase 2  | MIM, Other species Gorilla gorilla                | UniProtKB–P23786 (CPT2_HUMAN)<br>NCBI Blast P                 |
| 182    | MT000149        | Carnitine O-acetyltransferase   | ER, MIM, Peroxisomes                              | UniProtKB–P43155 (CACP_HUMAN)                                 |
| 183    | MT000168        | Cytochrome P450, family 11, subfamily B, polypeptide 1  | MIM, Pan troglodytes                              | UniProtKB–P15538 (C11B1_HUMAN)                                |
| 184    | MT001587        | Dihydrofolate reductase-like 1  | Mitochondrion and cytoplasm                       | UniProtKB–P00374 (DYR_HUMAN)                                  |
| 185    | MT001494        | Dehydrogenase/reductase (SDR family) member 1   | ER  | UniProtKB–Q96LJ7 (DHRS1_HUMAN)                                |
| 186    | MT000186        | Dihydroliipoamide dehydrogenase   | MM, Nucleus, Flagellum                            | UniProtKB–P09622 (DLDH_HUMAN)                                 |
| 187    | MT001245        | Dual specificity phosphatase 21   | MIM, Nucleus, Cytoplasm                           | UniProtKB–Q9H596 (DUS21_HUMAN)                                |
| 188    | MT000231        | Folylpolyglutamate synthase   | MIM, MM, Cytoplasm                                | UniProtKB–Q05932 (FOLC_HUMAN)                                 |
| 189    | MT000259        | Glutamic-oxaloacetic transaminase 2, mitochondrial  | MM, PM  | UniProtKB–P00505 (AATM_HUMAN)                                 |
| 190    | MT000265        | Glutathione peroxidase 4  | MM, cytoplasm                                     | UniProtKB–P36969 (GPX4_HUMAN)                                 |
| 191    | MT001647        | Glutathione S-transferase kappa 1   | Peroxisomes                                       | UniProtKB–Q9Y2Q3 (GSTK1_HUMAN)<br>The Human protein ATLAS     |
| 192    | MT000280        | Holocytochrome c synthase   | MIM, PM   | UniProtKB–P53701 (CCHL_HUMAN)                                 |
| 193    | MT000687        | HECT and RLD domain containing E3 ubiquitin protein ligase 2  | Nucleus, cytoplasm, Centriole                     | UniProtKB–O95714 (HERC2_HUMAN)                                |
| 194    | MT000291        | 3-Hydroxymethyl-3-methylglutaryl-CoA lyase  | MM, Peroxisome                                    | UniProtKB–P35914 (HMGCL_HUMAN)                                |
| 195    | MT000283        | Huntingtin  | ER, Nucleus, Cytoplasm                            | UniProtKB–P42858 (HD_HUMAN)                                   |
| 196    | MT000334        | Lectin, galactoside-binding, soluble, 3   | Nucleus, cytoplasm, Extracellular region secreted | UniProtKB–P17931 (LEG3_HUMAN)                                 |



**Table 2** (continued)

| Sr. no | Mitoproteome ID | Gene/protein name  | Location   | Source   |
|--------|-----------------|--|--|--|
| 197    | MT000353        | Methyl CpG-binding protein 2   | Nucleus  | UniProtKB-P51608 (MECP2_HUMAN)                 |
| 198    | MT000365        | Mercaptopyruvate sulfurtransferase   | Mitochondrion, Cytoplasm, Synapse                | UniProtKB-P25325 (THTM_HUMAN)                  |
| 199    | MT001536        | OCLA domain containing 2   | Endosome   | UniProtKB-Q56VL3 (OCAD2_HUMAN)                 |
| 200    | MT000449        | Opioid receptor, $\delta$ 1  | PM   | UniProtKB-P41143 (OPRD_HUMAN)                  |
| 201    | MT000850        | Parkinson protein 7  | PM, ER, Nucleus, Mitochondria, Cytoplasm         | UniProtKB-Q99497 (PARK7_HUMAN)                 |
| 202    | MT000473        | Prohibitin   | Cytoplasm, PM, Nucleus and MIM                   | UniProtKB-P35232 (PHB_HUMAN)                   |
| 203    | MT000851        | Prohibitin 2   | MIM, Nucleus, Cytoplasm, PM                      | UniProtKB-Q99623 (PHB2_HUMAN)                  |
| 204    | MT001124        | Phosphotyrosine interaction domain containing 1  | Cytoplasm  | UniProtKB-Q7Z2X4 (PCLI1_HUMAN)                 |
| 205    | MT000480        | Phospholipase A2, group IVA (cytosolic, calcium-dependent)                                   | Golgi Apparatus, Nuclear envelope, Cytoplasm     | UniProtKB-P47712 (PA24A_HUMAN)                 |
| 206    | MT000818        | Peroxiredoxin 3  | Mitochondrion, Cytoplasm, Early endosome         | UniProtKB-P30048 (PRDX3_HUMAN)                 |
| 207    | MT000514        | Presenilin 1   | Golgi Apparatus, ER, PM, Early endosome          | UniProtKB-P49768 (PSN1_HUMAN)                  |
| 208    | MT000515        | Presenilin 2   | Golgi apparatus, ER                              | UniProtKB-P49810 (PSN2_HUMAN)                  |
| 209    | MT000610        | Thymidylate synthetase   | Mitochondrion, Cytoplasm, Nucleus                | UniProtKB-P04818 (TYSY_HUMAN)                  |
| 210    | MT000577        | SRC proto-oncogene, non-receptor tyrosine kinase   | PM, MIM, Nucleus, Cytoskeleton                   | UniProtKB-P12931 (SRC_HUMAN)                   |
| 211    | MT001544        | NADH dehydrogenase (ubiquinone) complex I, assembly factor 6                                 | MIM, Cytoplasm, Nucleus                          | UniProtKB-Q330K2 (NDUF6_HUMAN)                 |
| 212    | MT000591        | Tafazzin   | Cytoplasm, Mitochondrion                         | UniProtKB-Q16635 (TAZ_HUMAN)                   |
| 213    | MT001502        | Thioesterase superfamily member 4  | Mitochondrion, Cytoplasm, PM                     | UniProtKB-Q5T1C6 (THEM4_HUMAN)                 |
| 214    | MT000129        | Cytochrome c oxidase subunit IV isoform 1  | MIM, Other species <i>Pan paniscus</i>           | UniProtKB-P13073 (COX41_HUMAN)<br>NCBI Blast P |
| 215    | MT000136        | Cytochrome c oxidase subunit VIIa polypeptide 2 (liver)                                      | MIM, Other species <i>Klebsiella pneumoniae</i>  | UniProtKB-P14406 (CX7A2_HUMAN)<br>NCBI Blast P |
| 216    | MT000525        | Pyroline-5-carboxylate reductase 1   | Mitochondrion, Other species <i>Pongo abelii</i> | UniProtKB-P32322 (P5CR1_HUMAN)<br>NCBI Blast P |
| 217    | MT001553        | Reactive oxygen species modulator 1  | MIM, Other species <i>Camelus ferus</i>          | UniProtKB-P60602 (ROMO1_HUMAN)<br>NCBI Blast P |
| 218    | MT000619        | Ubiquinol-cytochrome c reductase core protein I  | MIM, Other species <i>Pan troglodytes</i>        | UniProtKB-P31930 (QCR1_HUMAN)<br>NCBI Blast P  |
| 219    | MT001532        | DnaJ (Hsp40) homolog, subfamily C, member 19   | MIM, Other species <i>Orcinus orca</i>           | UniProtKB-Q96DA6 (TIM14_HUMAN)<br>NCBI Blast P |
| 220    | MT000036        | Solute carrier family 25 (mitochondrial carrier; adenine nucleotide translocator), member 6  | MIM, Other species <i>Macaca fascicularis</i>    | UniProtKB-P12236 (ADT3_HUMAN)<br>NCBI Blast P  |
| 221    | MT000057        | ATP synthase, H <sup>+</sup> transporting, mitochondrial Fo complex, subunit C2 (subunit 9)  | Mitochondrion, Other species <i>Pan paniscus</i> | UniProtKB-Q06055 (AT5G2_HUMAN)<br>NCBI Blast P |
| 222    | MT001506        | Sideroflexin 2   | MIM, Other species <i>Gorilla gorilla</i>        | UniProtKB-Q96NB2 (SFXN2_HUMAN)<br>NCBI Blast P |
| 223    | MT001523        | NADH dehydrogenase (ubiquinone) 1 $\alpha$ subcomplex, 11, 14.7 kDa                          | MIM, Other species <i>Pan troglodytes</i>        | UniProtKB-Q86Y39 (NDUAB_HUMAN)<br>NCBI Blast P |
| 224    | MT001538        | Solute carrier family 9, subfamily B (NHA2, cation proton antiporter 2), member 2            | Endosome, MIM, PM                                | UniProtKB-Q86UD5 (SL9B2_HUMAN)<br>NCBI Blast P |
| 225    | MT000139        | Cytochrome c oxidase subunit VIIc  | MIM, Other species <i>Pan troglodytes</i>        | UniProtKB-P15954 (COX7C_HUMAN)<br>NCBI Blast P |
| 226    | MT001374        | Solute carrier family 25 (mitochondrial carrier; adenine nucleotide translocator), member 31 | PM, Mitochondrion                                | UniProtKB-Q9H0C2 (ADT4_HUMAN)<br>NCBI Blast P  |

**Table 2** (continued)

| Sr. no | Mitoproteome ID | Gene/protein name   | Location  | Source   |
|--------|-----------------|---|---|--|
| 227    | MT000226        | FGR proto-oncogene, Src family tyrosine kinase  | Cytosol, MIM, PM, Cytoskeleton                      | UniProtKB-P09769 (FGR_HUMAN)                   |
| 228    | MT001372        | Solute carrier family 25 (mitochondrial iron transporter), member 28                      | MIM, Other species <i>Macaca mulatta</i>            | UniProtKB-Q96A46 (MFRN2_HUMAN)<br>NCBI Blast P |
| 229    | MT000397        | NADH dehydrogenase (ubiquinone) 1 $\alpha$ subcomplex, 3, 9 kDa                           | MIM, Other species <i>Pan troglodytes</i>           | UniProtKB-O95167 (NDUA3_HUMAN)<br>NCBI Blast P |
| 230    | MT000399        | NADH dehydrogenase (ubiquinone) 1 $\alpha$ subcomplex, 5                                  | MIM, Other species <i>Pan troglodytes</i>           | UniProtKB-Q16718 (NDUA5_HUMAN)<br>NCBI Blast P |
| 231    | MT000400        | NADH dehydrogenase (ubiquinone) 1 $\alpha$ subcomplex, 6, 14 kDa                          | MIM, Other species <i>Pan troglodytes</i>           | UniProtKB-P56556 (NDUA6_HUMAN)<br>NCBI Blast P |
| 232    | MT000401        | NADH dehydrogenase (ubiquinone) 1 $\alpha$ subcomplex, 7, 14.5 kDa                        | MIM, Other species <i>Pan troglodytes</i>           | UniProtKB-O95182 (NDUA7_HUMAN)<br>NCBI Blast P |
| 233    | MT001234        | Proline dehydrogenase (oxidase) 2   | MM, Other species <i>Pan paniscus</i>               | UniProtKB-Q9UF12 (HYPDH_HUMAN)<br>NCBI Blast P |
| 234    | MT000406        | NADH dehydrogenase (ubiquinone) 1 $\beta$ subcomplex, 1, 7 kDa                            | MIM, Other species <i>Ostrea edulis</i>             | UniProtKB-O75438 (NDUB1_HUMAN)<br>NCBI Blast P |
| 235    | MT000408        | NADH dehydrogenase (ubiquinone) 1 $\beta$ subcomplex, 3, 12 kDa                           | MIM, Other species <i>Pan troglodytes</i>           | UniProtKB-O43676 (NDUB3_HUMAN)<br>NCBI Blast P |
| 236    | MT000409        | NADH dehydrogenase (ubiquinone) 1 $\beta$ subcomplex, 4, 15 kDa                           | MIM, Other species <i>Pan troglodytes</i>           | UniProtKB-O95168 (NDUB4_HUMAN)<br>NCBI Blast P |
| 237    | MT000410        | NADH dehydrogenase (ubiquinone) 1 $\beta$ subcomplex, 5, 16 kDa                           | MIM, Other species <i>Pan troglodytes</i>           | UniProtKB-O43674 (NDUB5_HUMAN)<br>NCBI Blast P |
| 238    | MT000411        | NADH dehydrogenase (ubiquinone) 1 $\beta$ subcomplex, 6, 17 kDa                           | MIM, Other species <i>Pan troglodytes</i>           | UniProtKB-O95139 (NDUB6_HUMAN)<br>NCBI Blast P |
| 239    | MT001186        | NADH dehydrogenase (ubiquinone) 1 $\alpha$ subcomplex, 12                                 | MIM, Other species <i>Pan troglodytes</i>           | UniProtKB-Q9UI09 (NDUAC_HUMAN)<br>NCBI Blast P |
| 240    | MT000417        | NADH dehydrogenase (ubiquinone) 1, subcomplex unknown, 2, 14.5 kDa                        | MIM, Other species <i>Gorilla gorilla</i>           | UniProtKB-O95298 (NDUC2_HUMAN)<br>NCBI Blast P |
| 241    | MT000422        | NADH dehydrogenase (ubiquinone) Fe-S protein 4, 18 kDa (NADH-coenzyme Q reductase)        | MIM, Other species <i>Gorilla gorilla</i>           | UniProtKB-O43181 (NDUS4_HUMAN)<br>NCBI Blast P |
| 242    | MT000423        | NADH dehydrogenase (ubiquinone) Fe-S protein 5, 15 kDa (NADH-coenzyme Q reductase)        | MIM, Other species <i>Pan troglodytes</i>           | UniProtKB-O43920 (NDUS5_HUMAN)<br>NCBI Blast P |
| 243    | MT000424        | NADH dehydrogenase (ubiquinone) Fe-S protein 6, 13 kDa (NADH-coenzyme Q reductase)        | MIM, Other species <i>Pan troglodytes</i>           | UniProtKB-O75380 (NDUS6_HUMAN)<br>NCBI Blast P |
| 244    | MT000425        | NADH dehydrogenase (ubiquinone) Fe-S protein 8, 23 kDa                                    | MIM, Other species <i>Pan troglodytes</i>           | UniProtKB-O00217 (NDUS8_HUMAN)<br>NCBI Blast P |
| 245    | MT000553        | Succinate dehydrogenase complex, subunit C, integral membrane protein, 15 kDa             | MM, Other species <i>Pan paniscus</i>               | UniProtKB-Q99643 (C560_HUMAN)<br>NCBI Blast P  |
| 246    | MT000798        | ATP synthase, H <sup>+</sup> transporting, mitochondrial Fo complex, subunit G            | MIM, Other species <i>Macaca mulatta</i>            | UniProtKB-O75964 (ATP5L_HUMAN)<br>NCBI Blast P |
| 247    | MT000950        | Translocase of inner mitochondrial membrane 10 homolog (yeast)                            | MIM, Other species <i>Macaca mulatta</i>            | UniProtKB-P62072 (TIM10_HUMAN)<br>NCBI Blast P |
| 248    | MT000692        | Solute carrier family 25 (mitochondrial carrier, brain), member 14                        | MIM, Other species <i>Macaca mulatta</i>            | UniProtKB-O95258 (UCP5_HUMAN)<br>NCBI Blast P  |
| 249    | MT000959        | ATP synthase, H <sup>+</sup> transporting, mitochondrial Fo complex, subunit s (factor B) | Mitochondrion, Other species <i>Pongo Abellii</i>   | UniProtKB-Q99766 (ATP5S_HUMAN)<br>NCBI Blast P |
| 250    | MT000722        | Chromosome 14 open reading frame 2  | Mitochondrion, Other species <i>Pan troglodytes</i> | UniProtKB-P56378 (ATP68_HUMAN)<br>NCBI Blast P |
| 251    | MT000728        | Solute carrier family 25, member 44   | Mitochondrion, Other species <i>Papio anubis</i>    | UniProtKB-Q96H78 (S2544_HUMAN)<br>NCBI Blast P |
| 252    | MT000756        | Solute carrier family 25 (aspartate/glutamate carrier), member 13                         | MIM, Other species <i>Pan paniscus</i>              | UniProtKB-Q9UJS0 (CMC2_HUMAN)<br>NCBI Blast P  |

**Table 2** (continued)

| Sr. no | Mitoproteome ID | Gene/protein name   | Location   | Source   |
|--------|-----------------|---|--|--|
| 253    | MT000757        | Solute carrier family 25 (mitochondrial carrier; ornithine transporter) member 15 | MIM, Other species <i>Cercocebus atys</i>                            | UniProtKB–Q9Y619 (ORNT1_HUMAN)<br>NCBI Blast P                           |
| 254    | MT001694        | Phospholipase A2, group IVB (cytosolic)   | Cytosol, Mitochondrion, Endosome                                     | UniProtKB–P0C869 (PA24B_HUMAN)<br>NCBI Blast P                           |
| 255    | MT000398        | NDUFA4, mitochondrial complex-associated  | MIM, Other species <i>Pan troglodytes</i>                            | UniProtKB–O00483 (NDUA4_HUMAN)   |
| 256    | MT000660        | <b>Nipsnap homolog 1 (<i>C. elegans</i>)</b>                                      | Mitochondrion and PM   | UniProtKB – Q9BPW8 (NIPS1_HUMAN)   |
| 257    | MT001369        | <b>Fumarylacetoacetate hydrolase domain containing 1</b>                          | Mitochondrion and Cytosol  | UniProtKB–Q6P587 (FAHD1_HUMAN)   |
| 258    | MT000921        | <b>NADH dehydrogenase (ubiquinone) complex I, assembly factor 3</b>               | MIM, Nucleus   | UniProtKB–Q9BU61 (NDUF3_HUMAN)   |
| 259    | MT000018        | Alanine-glyoxylate aminotransferase   | Mitochondrion, Peroxisomes   | UniProtKB–P21549 (SPYA_HUMAN)<br>The Human protein ATLAS                 |
| 260    | MT000741        | V-akt murine thymoma viral oncogene homolog 3                                     | Nucleus, Cytoplasm, PM   | UniProtKB–Q9Y243 (AKT3_HUMAN)  |
| 261    | MT000073        | Branched chain amino-acid transaminase 2, mitochondrial                           | Mitochondrion, Cytoplasm   | UniProtKB–O15382 (BCAT2_HUMAN)<br>The Human protein ATLAS                |
| 262    | MT000093        | Biotinidase   | Extracellular space  | UniProtKB–P43251 (BTD_HUMAN)<br>The Human protein ATLAS                  |
| 263    | MT000095        | Complement component 1, q subcomponent-binding protein                            | PM, Nucleus, Cytoplasm, MM   | UniProtKB–Q07021 (C1QBP_HUMAN)<br>The Human protein ATLAS                |
| 264    | MT000108        | Calsequestrin 1 (fast-twitch, skeletal muscle)                                    | ER, MM, Sarcoplasmic reticulum                                       | UniProtKB–P31415 (CASQ1_HUMAN)<br>The Human protein ATLAS                |
| 265    | MT001230        | Cell cycle and apoptosis regulator 2  | Nucleus, Cytoplasm, Cytoskeleton                                     | UniProtKB–Q8N163 (CCAR2_HUMAN)<br>The Human protein ATLAS                |
| 266    | MT000187        | Dihydrolipoamide S-succinyltransferase (E2 component of 2-oxo-glutarate complex)  | MM, Nucleus  | UniProtKB–P36957 (ODO2_HUMAN)  |
| 267    | MT000190        | DNA replication helicase/nuclease 2   | Mitochondrion, Nucleus   | UniProtKB–P51530 (DNA2_HUMAN)  |
| 268    | MT000695        | DnaJ (Hsp40) homolog, subfamily A, member 3                                       | PM, Mitochondrion, Cytosol, Nucleus                                  | UniProtKB–Q96EY1 (DNJA3_HUMAN)<br>The Human protein ATLAS                |
| 269    | MT000209        | Erb-b2 receptor tyrosine kinase 4   | PM, Mitochondrion, Nucleus   | UniProtKB–Q15303 (ERBB4_HUMAN)   |
| 270    | MT000891        | Ethylmalonic encephalopathy 1   | MM, Nucleus, Cytoplasm   | UniProtKB–O95571 (ETHE1_HUMAN)<br>The Human protein ATLAS                |
| 271    | MT000227        | Fumarate hydratase  | Mitochondrion, Nucleus, Cytosol                                      | UniProtKB–P07954 (FUMH_HUMAN)<br>The Human protein ATLAS                 |
| 272    | MT000237        | Glycyl-tRNA synthetase  | Cytoplasm, Extracellular region, Mitochondrion                       | UniProtKB–P41250 (GARS_HUMAN)<br>The Human protein ATLAS                 |
| 273    | MT000253        | Glutaminase   | Cytosol, MM, Cytoplasm   | UniProtKB–O94925 (GLSK_HUMAN)<br>The Human protein ATLAS                 |
| 274    | MT000271        | Glutathione reductase   | Mitochondrion, Cytoplasm   | UniProtKB–P00390 (GSHR_HUMAN)<br>The Human protein ATLAS                 |
| 275    | MT000276        | Hydroxyacylglutathione hydrolase  | MM, Cytoplasm  | UniProtKB–Q16775 (GLO2_HUMAN)  |
| 276    | MT000299        | Heat shock 70 kDa protein 1-like  | PM, MM, Nucleus, Cytosol   | UniProtKB–P34931 (HS71L_HUMAN)<br>The Human protein ATLAS                |
| 277    | MT000892        | Iron sulfur cluster assembly enzyme   | Cytoplasm, Mitochondrion, Nucleus, other species <i>Pongo abelii</i> | UniProtKB–Q9H1K1 (ISCU_HUMAN)<br>NCBI Blast P<br>The Human protein ATLAS |
| 278    | MT000326        | Lysyl-tRNA synthetase   | PM, Nucleus, Cytoplasm, Extracellular region                         | UniProtKB–Q15046 (SYK_HUMAN)<br>The Human protein ATLAS                  |
| 279    | MT000354        | Myocyte enhancer factor 2A  | Cytoplasm, Nucleoplasm, Mitochondrion                                | UniProtKB–Q02078 (MEF2A_HUMAN)   |
| 280    | MT000885        | Malonyl-CoA decarboxylase   | MM, Cytoplasm, Peroxisome  | UniProtKB–O95822 (DCMC_HUMAN)  |
| 281    | MT001351        | Mitochondrial transcription termination factor 2                                  | Mitochondrion, Nucleoid  | UniProtKB–Q49AM1 (MTEF2_HUMAN)   |

**Table 2** (continued)

| Sr. no | Mitoproteome ID | Gene/protein name   | Location  | Source   |
|--------|-----------------|---|---|--|
| 282    | MT001530        | Mitochondrial transcription termination factor 4                            | Mitochondrion, Chloroplast and cytosol              | UniProtKB-Q9ZT96 (MTEF4_ARATH)                           |
| 283    | MT000267        | Nuclear receptor subfamily 3, group C, member 1 (glucocorticoid receptor)   | Cytoplasm, Cytoskeleton, Nucleus, Mitochondrion     | UniProtKB-P04150 (GCR_HUMAN)                             |
| 284    | MT000375        | Nudix (nucleoside diphosphate linked moiety X)-type motif 1                 | Cytosol, MM, Nucleus                                | UniProtKB-P36639 (8ODP_HUMAN)<br>The Human protein ATLAS |
| 285    | MT000446        | Oxoglutarate ( $\alpha$ -ketoglutarate) dehydrogenase (lipoamide)           | Nucleus and MM                                      | UniProtKB-Q02218 (ODO1_HUMAN)                            |
| 286    | MT000465        | Pyruvate dehydrogenase kinase, isozyme 1                                    | Nucleus, Cytoplasm, PM, MM                          | UniProtKB-O15530 (PDPK1_HUMAN)                           |
| 287    | MT001341        | PIF1 5'-to-3' DNA helicase  | Nucleus and mitochondrion                           | UniProtKB-Q9H611 (PIF1_HUMAN)                            |
| 288    | MT000477        | Protein (peptidylprolyl cis/trans isomerase) NIMA-interacting, 4 (parvulin) | Nucleus, Cytoplasm, MM, Cytoskeleton                | UniProtKB-Q9Y237 (PIN4_HUMAN)                            |
| 289    | MT000484        | Polymerase (DNA directed), $\gamma$   | Mitochondrial, Nucleoid                             | UniProtKB-P54098 (DPOG1_HUMAN)                           |
| 290    | MT000455        | Peroxiredoxin 1   | Cytoplasm, Melanosome                               | UniProtKB-Q06830 (PRDX1_HUMAN)                           |
| 291    | MT000915        | Peroxiredoxin 5   | Cytosol, Mitochondrion, Peroxisome matrix           | UniProtKB-P30044 (PRDX5_HUMAN)                           |
| 292    | MT001591        | Primase and polymerase (DNA-directed)                                       | Nucleus, MM, Chromosome                             | UniProtKB-Q96LW4 (PRIPO_HUMAN)                           |
| 293    | MT000527        | Glutaminyl-tRNA synthetase  | Cytosol, Cytoplasm                                  | UniProtKB-P47897 (SYQ_HUMAN)                             |
| 294    | MT001602        | Ribonuclease H1   | Cytoplasm   | UniProtKB-O60930 (RNH1_HUMAN)                            |
| 295    | MT001002        | Ribonucleotide reductase M2 B (TP53 inducible)                              | Nucleus, Cytoplasm                                  | UniProtKB-Q7LG56 (RIR2B_HUMAN)                           |
| 296    | MT001379        | Sestrin 2   | Cytoplasm   | UniProtKB-P58004 (SESN2_HUMAN)                           |
| 297    | MT001371        | SRA stem-loop-interacting RNA-binding protein                               | Mitochondrion, Nucleus                              | UniProtKB-Q9GZT3 (SLIRP_HUMAN)                           |
| 298    | MT000886        | Tudor domain containing 7   | Cytoplasm   | UniProtKB-Q8NHU6 (TDRD7_HUMAN)                           |
| 299    | MT001226        | tRNA methyltransferase 5  | MM, Nucleus, Cytoplasm                              | UniProtKB-Q32P41 (TRM5_HUMAN)                            |
| 300    | MT000197        | Thymidine phosphorylase   | Cytosol   | UniProtKB-P19971 (TYPH_HUMAN)                            |
| 301    | MT001350        | GrpE-like 1, mitochondrial (E. coli)  | MM, Other species <i>Pongo abelii</i>               | UniProtKB-Q9HAV7 (GRPE1_HUMAN)<br>NCBI Blast P           |
| 302    | MT001284        | Mitochondrial ribosomal protein L34   | Mitochondrion, Other species <i>Pongo abelii</i>    | UniProtKB-Q9BQ48 (RM34_HUMAN)<br>NCBI Blast P            |
| 303    | MT000405        | NADH dehydrogenase (ubiquinone) 1, $\alpha/\beta$ subcomplex, 1, 8 kDa      | Mitochondrion, Other species <i>Pan troglodytes</i> | UniProtKB-O14561 (ACPM_HUMAN)<br>NCBI Blast P            |
| 304    | MT000462        | Pyruvate dehydrogenase (lipoamide) $\alpha$ 1                               | MM, Other species <i>Theropithecus gelada</i>       | UniProtKB-P08559 (ODPA_HUMAN)<br>NCBI Blast P            |
| 305    | MT000466        | Pyruvate dehydrogenase kinase, isozyme 2                                    | MM, Other species <i>Pan troglodytes</i>            | UniProtKB-Q15119 (PDK2_HUMAN)<br>NCBI Blast P            |

ER endoplasmic reticulum, PM plasma membrane, OM outer membrane, MM mitochondrial matrix, IMS inter-membrane space, MIM mitochondrial inner membrane

### To the Editor, The Mitoproteome database

Mitoproteome, human mitochondrial protein database is freely available at <http://www.mitoproteome.org/>. It contains known mitochondrial proteins obtained either from mass spectrometric analysis of highly purified human heart mitochondria or retrieved from other publicly available databases (Cotter 2004). Other human mitochondrial protein databases do not have the complete information about all mitochondrial and nuclear-encoded proteins (Ravichandran et al. 2016). Databases

dealing with mitochondrial proteins primarily highlight mitochondrial and nuclear-encoded proteins and localization in mitochondria.

### Whether the Mitoproteome database represents exclusively mitochondrial proteins?

In the Mitoproteome database, we have observed that some of the mentioned proteins were exclusively mitochondrial, while others can be found at other organelles also. In this context, we had searched the literature and

various databases like The Human protein Atlas, UniProt, HGNC database and OMIM database to classify the proteins on the aforementioned basis and made a separate table for exclusively mitochondrial and non-mitochondrial proteins/genes. The NCBI Protein Blast was used to sort mitochondrial proteins/genes that showed 100% sequence similarity to humans and were listed as exclusive mitochondrial proteins/genes of humans (Table 1). Some common examples of exclusive human mitochondrial proteins are citrate synthase, superoxide dismutase 2, succinate dehydrogenase complex, etc. The remaining proteins/genes that show sequence similarity to proteins of other organelles are listed as non-exclusive mitochondrial genes/proteins (Table 2). Examples of non-exclusive mitochondrial proteins are fumarate hydratase, glutathione reductase, sestrin 2, etc.

In some situations, knowledge of exclusive proteins is crucial. For example, cyanide is lethal as it inhibits cytochrome c oxidase, a mitochondrial ETC protein. It inhibits an exclusive mitochondrial function needed for maintenance of life process. We are continuously getting exposed to several chemicals, and such exposure is definitely having effect on mitochondrial function. As an example, aluminum phosphide (rice or wheat pill) which induces phosphine exposure acts as mitochondrial poison (Yadav et al. 2021). We do not know entirely with which mitochondrial proteins it interacts. This is because we do not have complete information about exclusive mitochondrial proteins. We believe that data generated here will pave the path in such direction. Further a protein whose information is mentioned in a mitochondrial database so far generated is not always an exclusive mitochondrial protein, but it can be misunderstood so from the name of the database. For example (Table 2, Sr. No.2), acyl-CoA synthetase long-chain family member 1 is a protein whose information is recorded in Mitoproteome database; but it is also found in peroxisomes. So, the information generated in the present study will help the researchers to get rid of such confusion.

Sorting of exclusive mitochondrial proteins is necessary because these interaction networks have various roles in maintaining the normal physiology of cells (Pourahmad et al. 2015). Dysfunction of mitochondrial proteins/genes has been associated with neurological, liver, oxygen deficiency, inborn and metabolic diseases (Javadov et al. 2020). Therefore, knowledge about mitochondrial proteins/genes is essential to understand disease progression, mitochondrial evolution, functional protein-protein interaction network, cancer prevention, stem cell and regeneration biology (Chakrabarty et al. 2018). We believe that if such information is available with the Mitoproteome database, it will help the researchers.

## Conclusions

This work documents the list of exclusively mitochondrial proteins known as on date. With the progress of human knowledge, the list needs to be updated and revised.

## Abbreviations

MTS: Mitochondrial targeting sequence; TOM: Translocase of outer membrane; CLUH: Clustered mitochondria homolog.

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## Author contributions

DY has done the sorting work under the supervision of DB and RB. DY wrote the primary manuscript. BT was present throughout the study, and she cross-checked the results. DK and SC were associated with the planning phase of the study. They also verified the results and cross-checked the references. All the authors approved the final version of the manuscript.

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## Availability of data and materials

The datasets generated and/or analyzed during the current study are available at <http://www.mitoproteome.org/>, <https://www.uniprot.org/>, <http://www.ncbi.nlm.nih.gov/gene/8165>.

## Declarations

### Ethical approval and consent to participate

This is a computer-based analysis. Hence, no ethical approval is required.

### Consent for publication

This work does not require any type of sample. Hence, informed consent is not required.

### Competing interests

The authors declare that they have no competing interests.

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