

**Table S4:** Differentially regulated mitochondrial proteins in the PS of *rod* <sup>$\Delta$ Vhl</sup> mice

| Uniprot ID | Gene symbol | Abundance Ratio<br>[ <i>rod</i> <sup><math>\Delta</math>Vhl</sup> / ctrl] | P Value   |
|------------|-------------|---------------------------------------------------------------------------|-----------|
| O55003     | Bnip3       | 100                                                                       | <0.000001 |
| Q3V3R1     | Mthfd1l     | 4.04                                                                      | <0.000001 |
| O35143     | ATP5F1      | 3.04                                                                      | <0.000001 |
| Q922F7     | Bnip3l      | 2.08                                                                      | 0.004     |
| Q922H2     | Pdk3        | 1.48                                                                      | 0.013     |
| P09528     | Fth1        | 1.41                                                                      | 0.014     |
| Q8R4N0     | Clybl       | 1.39                                                                      | 0.047     |
| Q9D6U8     | Fam162a     | 1.37                                                                      | 0.024     |
| Q91Z53     | Grhpr       | 1.31                                                                      | 0.034     |
| Q9DC29     | Abcb6       | 0.80                                                                      | 0.042     |
| Q925I1     | Atad3       | 0.78                                                                      | 0.029     |
| Q99JB2     | Stoml2      | 0.77                                                                      | 0.040     |
| Q9CQZ6     | Ndufb3      | 0.77                                                                      | 0.022     |
| Q91VN4     | Chchd6      | 0.76                                                                      | 0.024     |
| P99029     | Prdx5       | 0.76                                                                      | 0.043     |
| Q8CIM3     | D2hgdh      | 0.74                                                                      | 0.039     |
| P52503     | Ndufs6      | 0.74                                                                      | 0.012     |
| Q921H8     | Acaa1a      | 0.74                                                                      | 0.006     |
| Q9D0M3     | Cyc1        | 0.73                                                                      | 0.024     |
| Q8BK08     | Tmem11      | 0.73                                                                      | 0.024     |
| Q9CR21     | Ndufab1     | 0.73                                                                      | 0.029     |
| Q9CZU6     | Cs          | 0.73                                                                      | 0.022     |
| Q9CQJ8     | Ndufb9      | 0.73                                                                      | 0.007     |
| P47740     | Aldh3a2     | 0.72                                                                      | 0.006     |
| Q9CRB9     | Chchd3      | 0.72                                                                      | 0.010     |
| Q9DCS3     | Mecr        | 0.71                                                                      | 0.004     |
| Q9DCS9     | Ndufb10     | 0.71                                                                      | 0.005     |
| Q99L13     | Hibadh      | 0.71                                                                      | 0.011     |
| P00405     | Mtco2       | 0.70                                                                      | 0.009     |
| Q9CYK1     | Wars2       | 0.70                                                                      | 0.042     |
| Q8C163     | Exog        | 0.69                                                                      | 0.012     |
| Q9WTP6     | Ak2         | 0.69                                                                      | 0.003     |
| Q9JLZ3     | Auh         | 0.68                                                                      | 0.006     |
| Q9WTP7     | Ak3         | 0.68                                                                      | 0.003     |
| Q9CQ69     | Uqcrcq      | 0.68                                                                      | 0.002     |
| Q9CPQ8     | Atp5mg      | 0.68                                                                      | 0.002     |
| Q66GT5     | Ptpmt1      | 0.68                                                                      | 0.006     |
| P12787     | Cox5a       | 0.68                                                                      | 0.004     |
| Q8BWM0     | Ptges2      | 0.67                                                                      | 0.001     |
| P97493     | Txn2        | 0.66                                                                      | 0.004     |
| P19783     | Cox4i1      | 0.66                                                                      | 0.002     |
| Q91ZE0     | Tmlhe       | 0.65                                                                      | 0.0005    |
| Q3UUI3     | Them4       | 0.65                                                                      | 0.002     |
| Q99LP6     | Grpel1      | 0.65                                                                      | 0.002     |
| P56380     | Nudt2       | 0.65                                                                      | 0.016     |
| Q924T2     | Mrps2       | 0.64                                                                      | 0.026     |
| Q9CPQ1     | Cox6c       | 0.63                                                                      | 0.0001    |
| Q9CQ75     | Ndufa2      | 0.63                                                                      | 0.001     |
| Q9QZM2     | Polg2       | 0.62                                                                      | 0.007     |
| P62897     | Cycs        | 0.62                                                                      | 0.0005    |
| Q9CQQ7     | Atp5pb      | 0.62                                                                      | 0.0004    |
| Q9CQ54     | Ndufc2      | 0.61                                                                      | 0.001     |
| Q9DCT2     | Ndufs3      | 0.60                                                                      | 0.0002    |
| Q9CPP6     | Ndufa5      | 0.60                                                                      | 0.0004    |
| Q9CQC7     | Ndufb4      | 0.59                                                                      | 0.001     |
| Q8BHC4     | Dcakd       | 0.59                                                                      | 0.001     |
| Q9CQZ5     | Ndufa6      | 0.58                                                                      | 0.000004  |
| P56135     | Atp5mf      | 0.58                                                                      | 0.00004   |
| Q9DB20     | Atp5po      | 0.57                                                                      | 0.00003   |
| P20108     | Prdx3       | 0.57                                                                      | 0.00003   |
| Q9DCJ5     | Ndufa8      | 0.56                                                                      | 0.000002  |
| Q8R2Y8     | Pthr2       | 0.55                                                                      | 0.0001    |
| Q9D1H6     | Ndufaf4     | 0.55                                                                      | 0.004     |
| Q9Z1P6     | Ndufa7      | 0.54                                                                      | 0.00005   |
| Q8R1I1     | Uqcrc10     | 0.54                                                                      | 0.00003   |
| P85094     | Isoc2a      | 0.50                                                                      | 0.000003  |
| Q9DCX2     | Atp5pd      | 0.49                                                                      | <0.000001 |
| Q9WVL0     | Gstz1       | 0.42                                                                      | <0.000001 |
| P99028     | Uqcrcr      | 0.39                                                                      | 0.00005   |
| Q9DCU6     | Mrpl4       | 0.36                                                                      | 0.00008   |
| O35943     | Fxn         | 0.01                                                                      | <0.000001 |
| P58044     | Idi1        | 0.01                                                                      | <0.000001 |
| Q9D6S7     | Mrrf        | 0.01                                                                      | <0.000001 |