

Supplementary Materials:

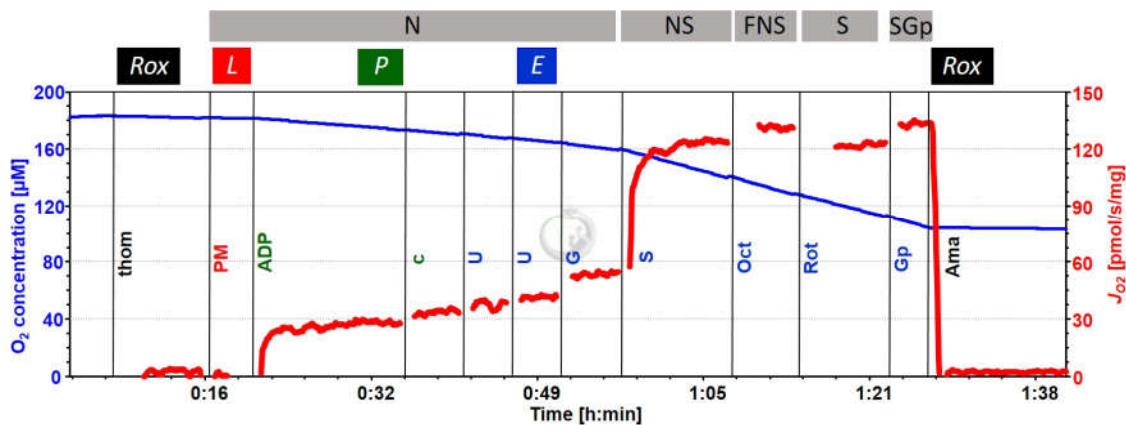


Figure S1. Representative trace of substrate-uncoupler-inhibitor titration (SUIT) protocol for measurement of liver mitochondrial respiration using high-resolution respirometry. Residual oxygen consumption (*Rox*) measured in the presence of liver homogenate (*thom*); pyruvate and malate (*PM*) in the absence of adenosine diphosphate (*ADP*) to measure NADH-linked (*N*) LEAK-respiration (*L*); kinetically saturating concentration of *ADP* to measure OXPHOS-capacity (*P*); cytochrome *c* (*c*) to detect mitochondrial outer membrane integrity; uncoupler titrations (*U*) to measure electron transfer (*ET*) capacity (*E*); glutamate (*G*) to measure *N*-linked *ET* capacity; succinate (*S*) to measure *NS*-linked *ET* capacity; octanoylcarnitine (*Oct*) to detect *FNS*-linked *ET* capacity; Complex I inhibitor rotenone (*Rot*) to measure *S*-*ET* capacity; glycerol-3-phosphate (*Gp*) to measure *SGp*-*ET* capacity; antimycin A (*Ama*) to detect *Rox*. Experiment 2021-02-01 PS1-01 WT ND 1: O_2 concentration (blue trace) and O_2 flux per mass (red trace).

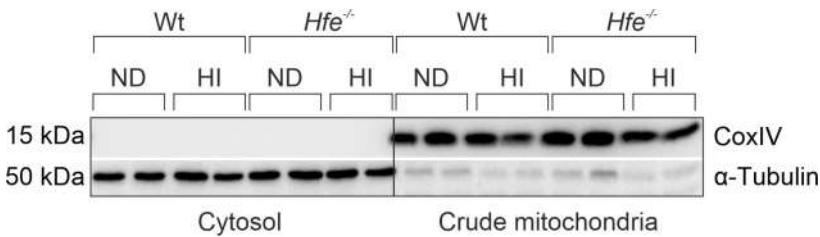


Figure S2. Representative Western blot of isolated liver fractions to ensure their purity. Mitochondrial protein cytochrome *c* oxidase subunit 4 (*CoxIV*) and cytosolic protein α -Tubulin.