

Fig. S1, related to figure 1: Sequence validation of VCP CRISPR knock-in disease models. Sequencing chromatograms for the genotypes indicated. Mutants are heterozygous; thus, in some cases two peaks are observed for the WT and mutant sequence.

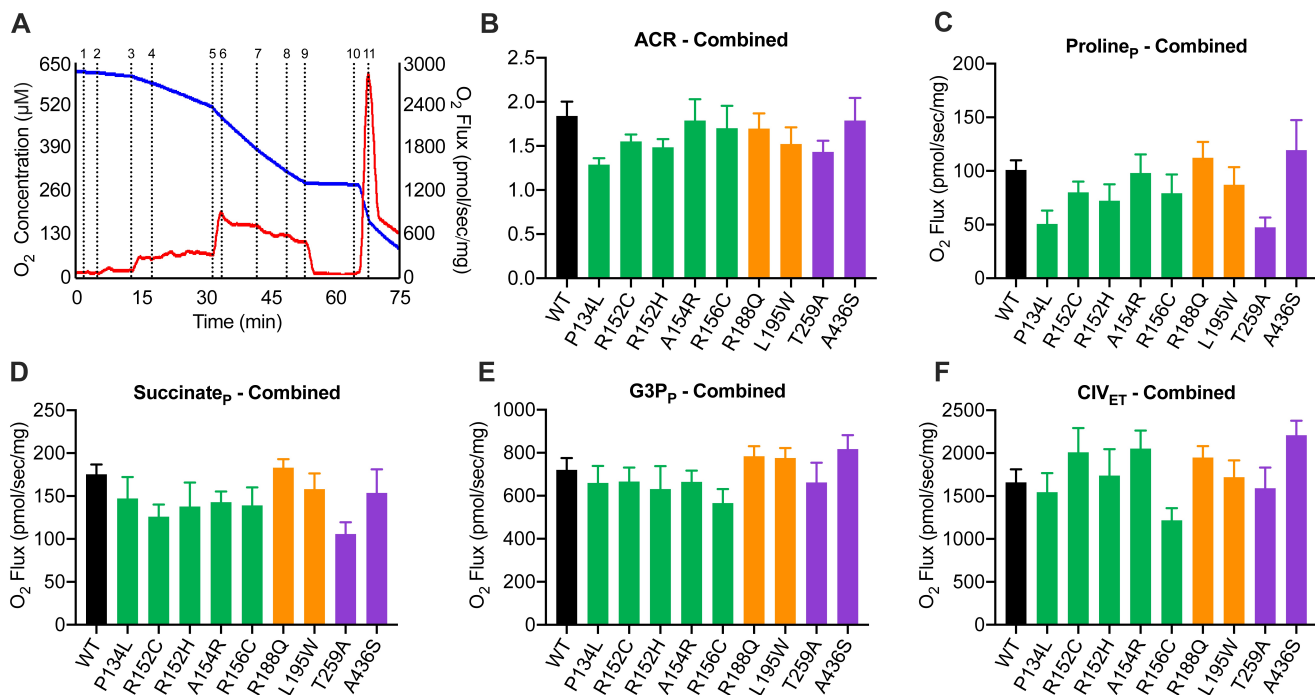


Fig. S2, related to Figure 8: Mitochondrial respiratory function in VCP mutants

A. Representative tracing of the change in oxygen concentration (blue) and flux (red) over the course of a substrate-uncoupler-inhibitor titration protocol. Numbers correspond with the following titrations: (1) pyruvate + malate, (2) ADP, (3) proline, (4) succinate, (5) glycerol-3-phosphate (G3P), (6) FCCP, (7) rotenone, (8) atpenin A5, (9) antimycin A, (10) ascorbate + TMPD, and (11) sodium azide. **B-F.** ACR (B) and respiration supported by proline (C), succinate (D), and G3P (E) in the presence of ADP (OXPHOS; P) and ascorbate + TMPD (CIV) (F) in aged male and female WT and VCP mutants. Data are presented as the mean \pm SEM and were assessed by one-way ANOVA with Dunnett's multiple comparisons.