

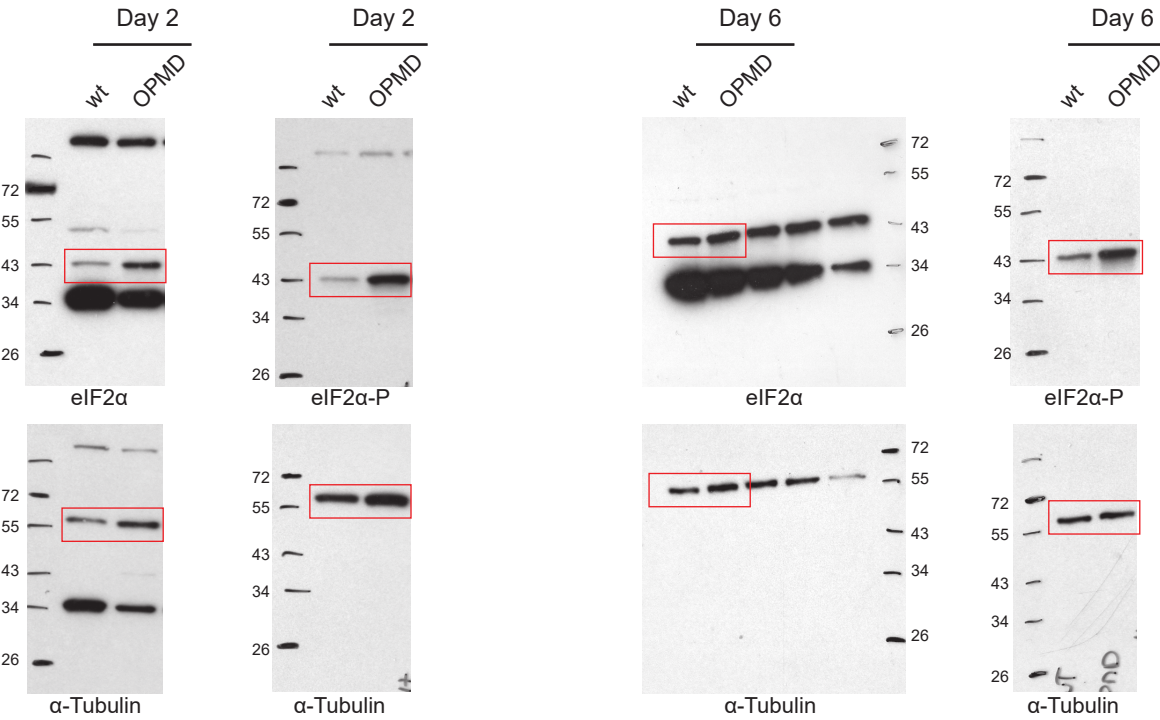
The small compound Icerguastat reduces muscle defects in oculopharyngeal muscular dystrophy through the PERK pathway of the unfolded protein response

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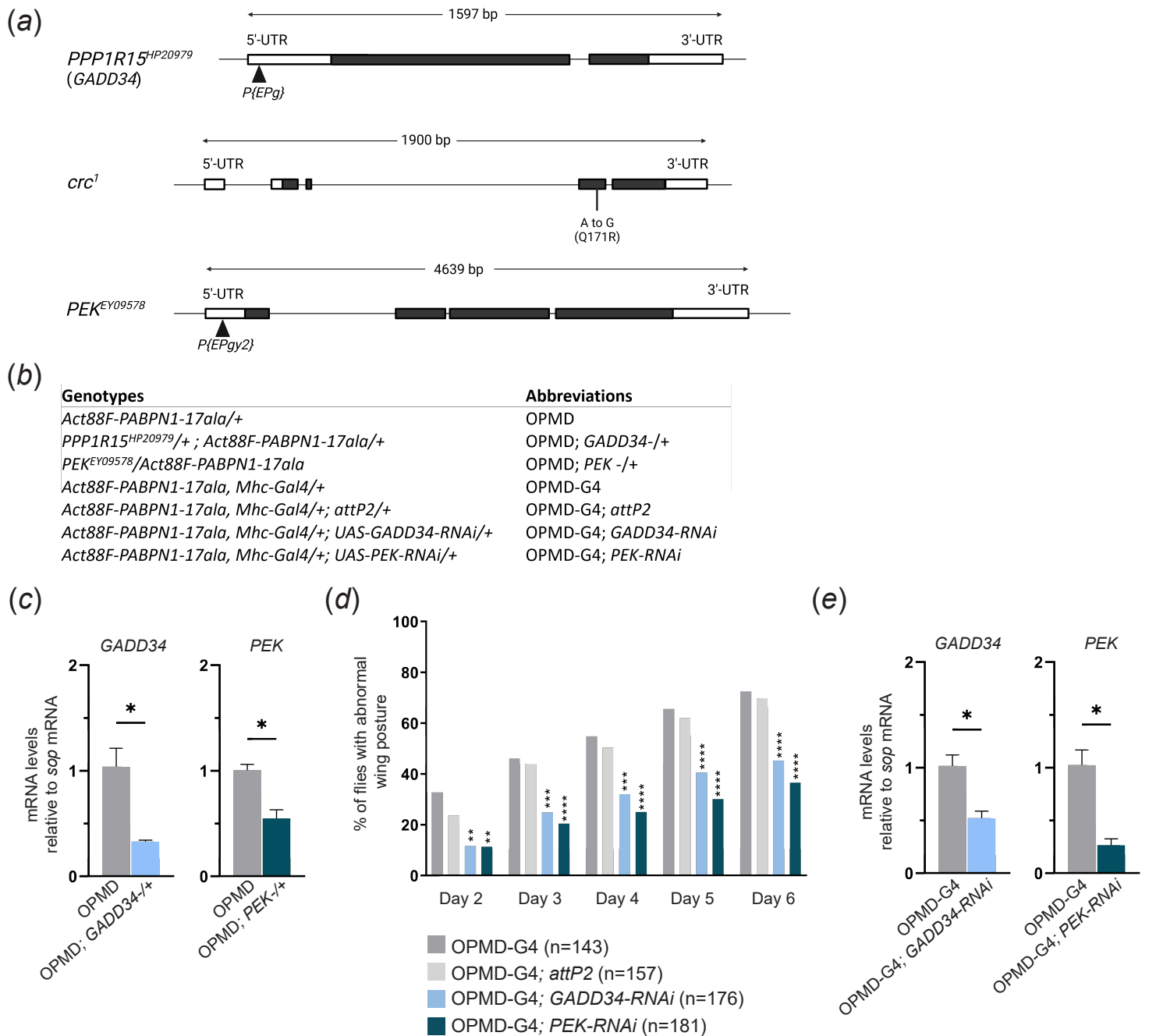
Supplementary material

The Supplementary material contains four supplementary figures.

(Related to Figure 1)



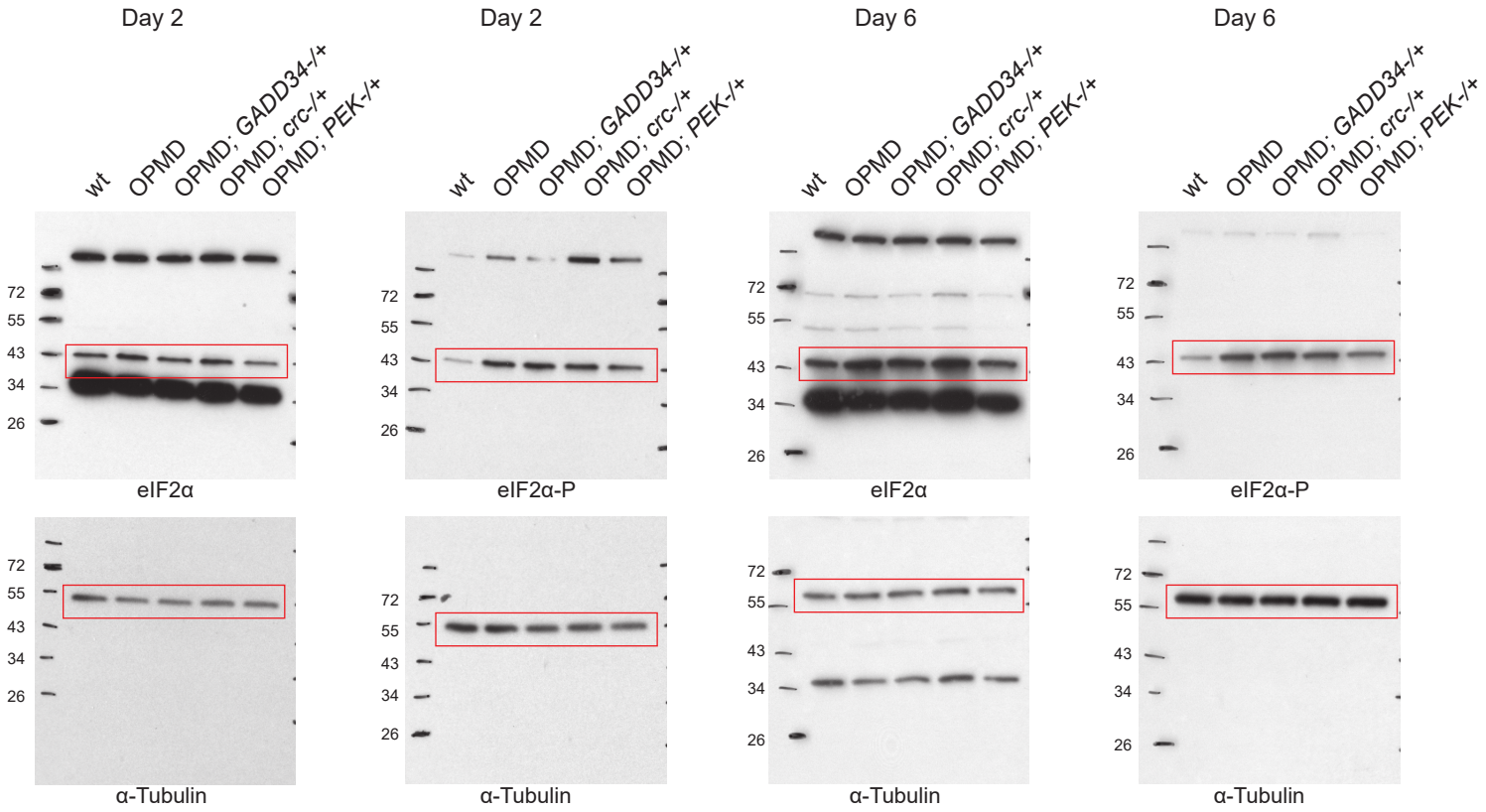
Supplementary Figure 1. Complete western blots for figure 1d.



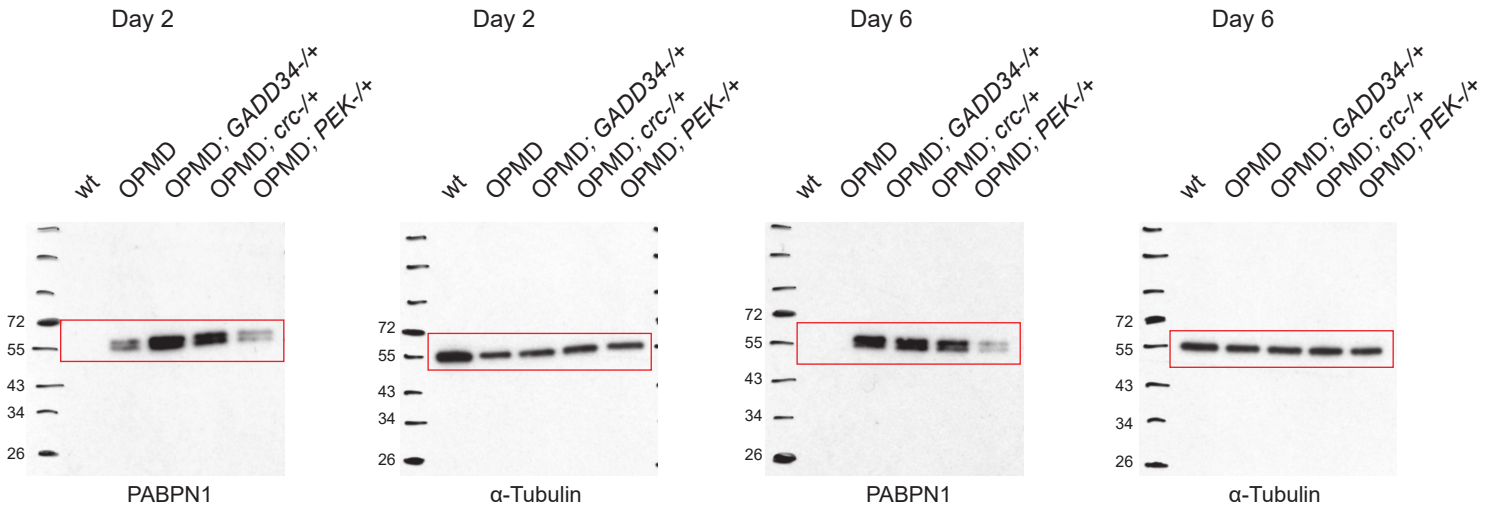
Supplementary Figure 2. Characterization of PERK pathway mutants.

(a) *GADD34* (*PPP1R15*) and *PEK* mutants have *P*-element insertions in their 5'-UTR. *crc* mutant is a hypomorphic allele with a nucleotide substitution [66]. Thin lines indicate intergenic regions and introns, open boxes represent UTRs and black boxes represent coding sequences. (b) Genotypes and abbreviations used in (c-e). (c) Quantification of *GADD34* and *PEK* mRNA levels in OPMD thoracic muscles in the absence or presence of the corresponding heterozygous mutant, at day 2, using RT-qPCR. mRNA levels were normalized to *sop* mRNA and set to 1 in OPMD thoraxes. Means are from four biological replicates quantified in triplicates, error bars represent standard deviations. * $p < 0.05$, using the unpaired-t test. (d) Wing position defects were scored in OPMD-G4 (*Act88F-PABPN1-17ala, Mhc-Gal4/+*) flies in the absence or presence of *GADD34* or *PEK* *UAS-RNAi* transgenes, each day between days 2 and 6. The numbers of scored flies are indicated (n). *attP2* is the landing site of *GADD34-UAS-RNAi* and *PEK-UAS-RNAi* and the *attP2* stock was used as a negative control. Quantifications were from two biological replicates. ** $p < 0.01$, *** $p < 0.001$, **** $p < 0.0001$, compared to OPMD-G4; *attP2*, using the Chi²-test. (e) Quantification of *GADD34* and *PEK* mRNA levels in OPMD thoracic muscles in the absence or presence of *GADD34-UAS-RNAi* or *PEK-UAS-RNAi* transgenes, respectively, at day 2 using RT-qPCR. mRNA levels were normalized to *sop* mRNA and set to 1 in OPMD-G4 thoraxes. Means are from four biological replicates quantified in triplicates, error bars represent standard deviations. * $p < 0.05$, using the unpaired-t test.

(a) (Related to Figure 2)

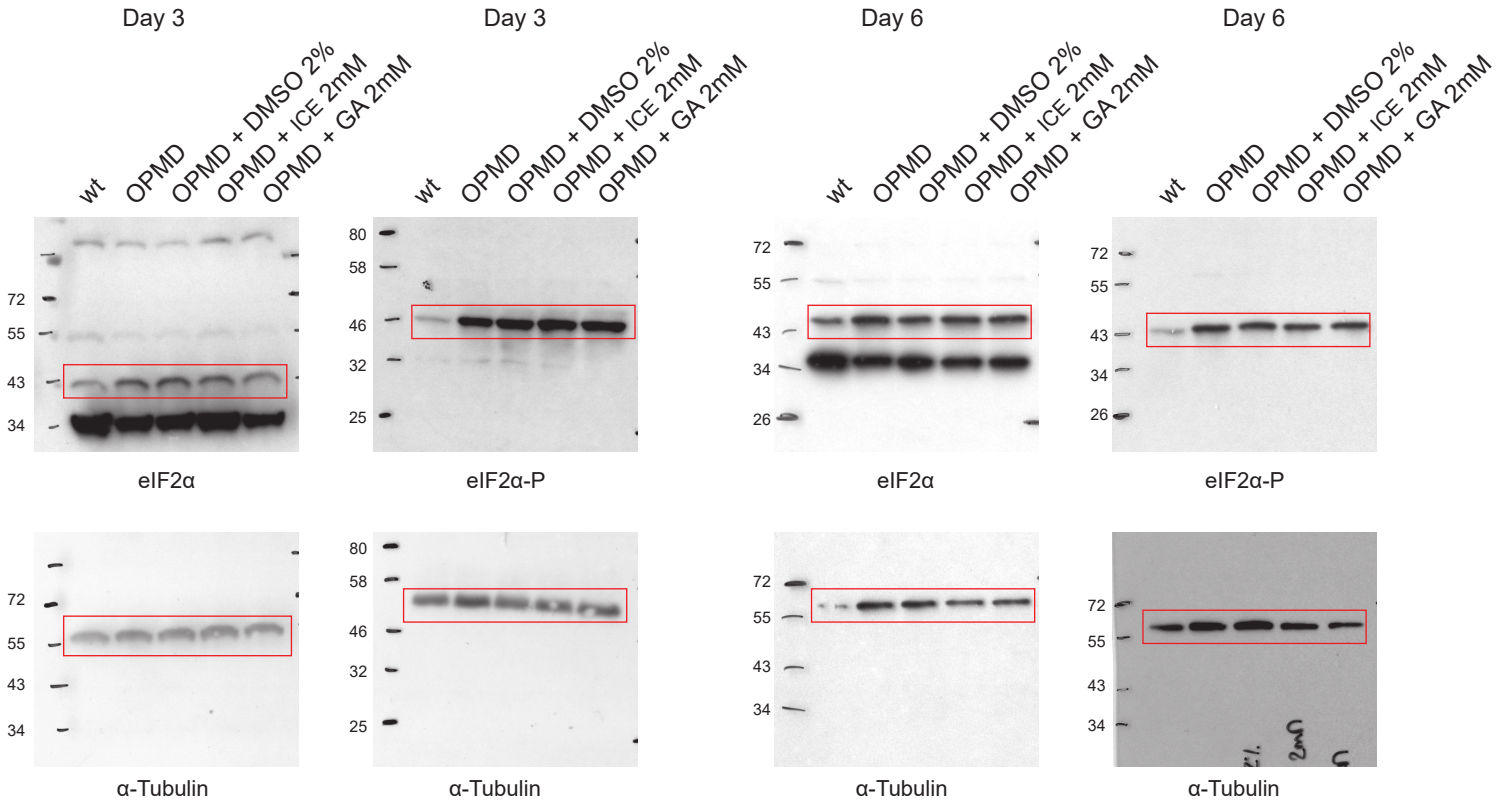


(b) (Related to Figure 3)

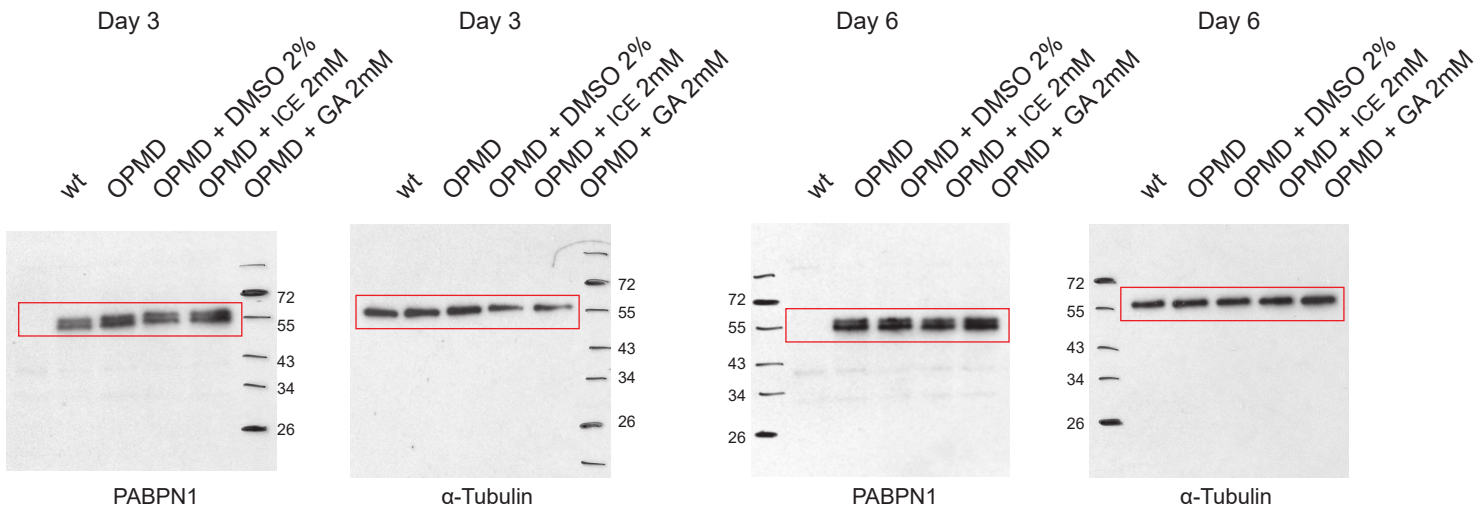


Supplementary Figure 3. Complete western blots for figure 2c (a) and figure 3e (b).

(a) (Related to Figure 4)



(b) (Related to Figure 6)



Supplementary Figure 4. Complete western blots for figure 4e (a) and figure 6e (b).