Supplementary Legends and Figures S1 and S2

Familial ALS-linked mutations in *Profilin 1* exacerbate TDP-43-induced degeneration in the retina of *Drosophila Melanogaster* through an increase in the cytoplasmic localization of TDP-43.

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Figure S1

Overexpression of fALS mutant PFN1 exacerbated TDP-43-induced retinal degeneration.

(A) Comparison of the expression levels of PFN1 (top panel), TDP-43 (middle panel) or α -tubulin (bottom panel) in 3 independent lines of each TG flies. Immunoblot analyses of the heads of 5-day-old *gmr*-driven TG flies expressing singly *LacZ* or *TDP-43*, or doubly *TDP-43* and *wt*, *C71G* or *M114T mt PFN1*. α -Tubulin levels are shown as a loading control (bottom panel). (B) Relative expression levels of PFN1 in (A) are shown. mean \pm *SE*. (C) H&E stained sections of eyes of 5-day-old *gmr*-driven TG flies overexpressing singly human *TDP-43*, or doubly *TDP-43* and *wt*, *C71G* or *M114T mutant PFN1*. Scale bar = 100 µm. (D) Retinal thickness measured in TG flies expressing singly *LacZ* or *TDP-43*, or doubly *TDP-43* and *wt*, *C71G* or *M114T mutant PFN1*. $N = 5 \sim 7$, mean $\pm SE$, (**); p < 0.01, (***); p < 0.001.

Figure S2

Overexpression of fALS mutant PFN1 did not affect FUS-induced retinal degeneration.

(A) External surface pictures of eyes of 5-day-old gmr-driven TG flies overexpressing singly human FUS, or doubly FUS and wt, C71G or M114T mutant PFN1. The enlarged view of each eye is shown in lower right corner. (B) H&E stained sections of eyes of 5-day-old gmr-driven TG flies singly FUS or doubly FUS and wt, C71G or M114T mutant PFN1. Scale bar = 100 μ m. (C) Retinal thickness measured in TG flies singly LacZ or FUS, or doubly FUS and wt, C71G or M114T mutant PFN1. N=10, mean \pm SE. (D) Comparison of the expression levels of FUS (upper panel) and PFN1 (middle panel). Immunoblot analyses of the heads of 5-day-old gmr-driven TG flies singly LacZ or FUS, or doubly FUS and wt, C71G or M114T mt PFN1. α -Tubulin levels are shown as a loading control (bottom panel). Relative expression levels of FUS are indicated under the panels. N=4, mean \pm SE.

Figure S1



