Supplementary Information

Late-stage rescue of visually guided behavior in the context of a significantly remodeled retinitis pigmentosa mouse model

Cellular and Molecular Life Sciences

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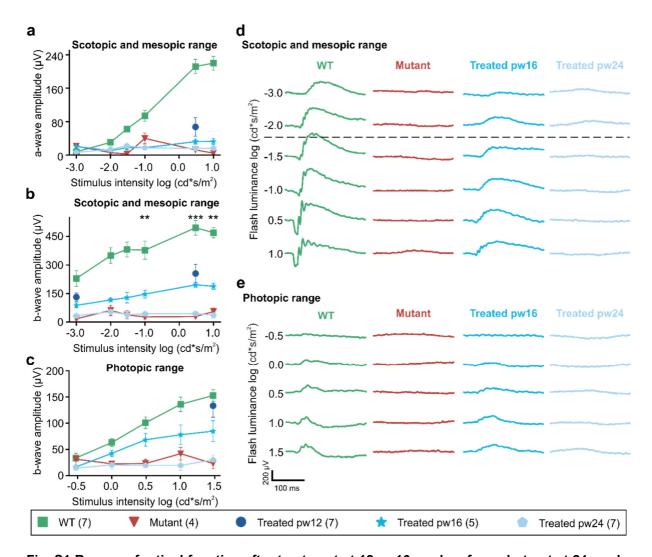


Fig. S1 Rescue of retinal function after treatment at 12 or 16 weeks of age, but not at 24 weeks $Pde6b^{STOP/STOP}$ mice were treated (or not) at pw12, pw16 or pw24, and full-field single-flash ERGs recorded at pw40. (a,b) Scotopic and mesopic a-wave and b-wave amplitudes. (c) Photopic b-wave amplitudes. Data represent the mean \pm SEM; Tukey's test for multiple comparisons. ** $P \le .01$; *** $P \le .001$. Asterisks, significant differences between mutant and treated at pw16. No statistical difference was found between animals treated at pw24 and mutant. N values, indicated in legend next to each group. Representative responses from a single retina in the (d) scotopic and mesopic range, and (e) in the photopic range. Dashed line, border between scotopic (above) and mesopic ranges (below).

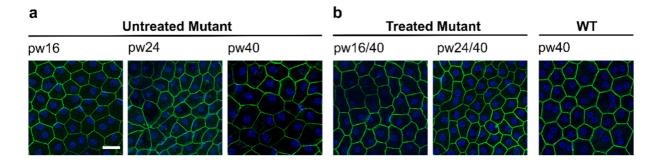


Fig. S2 Minor morphological abnormalities of the peripheral RPE $Pde6b^{STOP/STOP}$ mice were sacrificed at 16, 24 or 40 weeks of age. $Pde6b^{STOP/+}$ mice (WT) and $Pde6b^{STOP/STOP}$ mice treated at pw16 and pw24 were all sacrificed at pw40. RPE flat-mounts were labeled with an anti-β-catenin antibody (green) to visualize a component of adherens junctions. **(a)** Representative RPE flat-mounts from untreated $Pde6b^{STOP/STOP}$ mice at 16, 24 and 40 weeks of age and **(b)** from 40-week-old treated $Pde6b^{STOP/STOP}$ and $Pde6b^{STOP/+}$ (WT) mice. Scale bar, 20 μm.