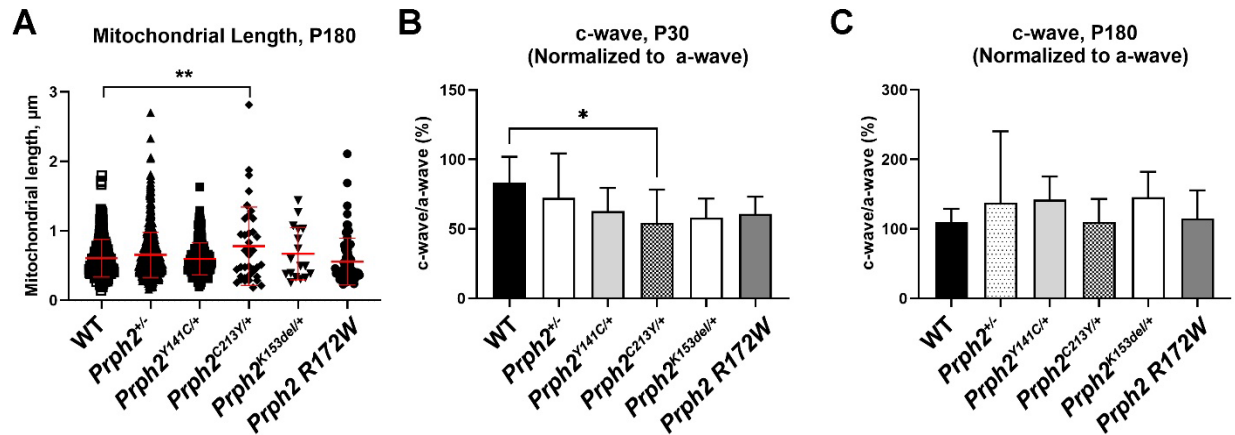
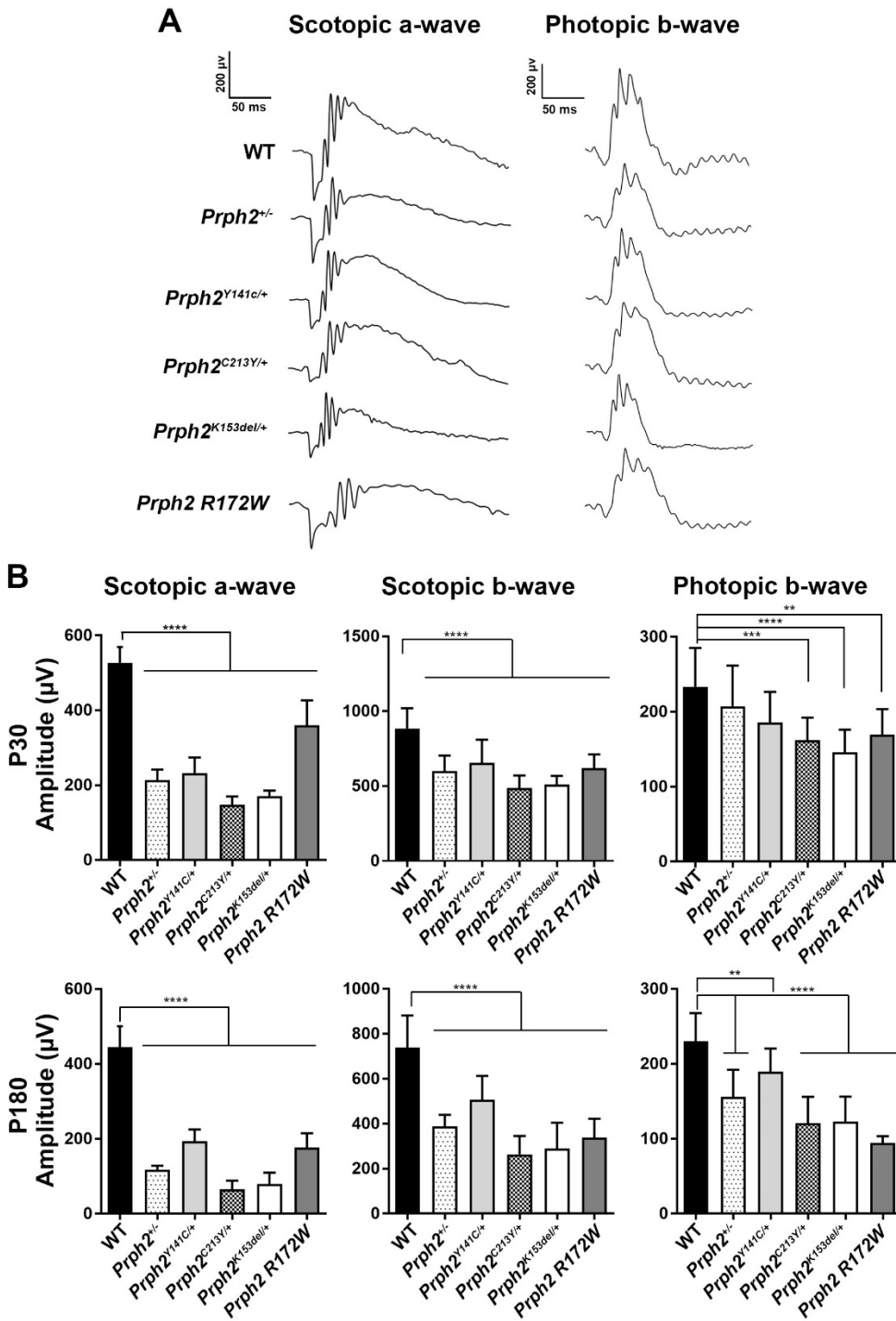


Supplemental Figure 1

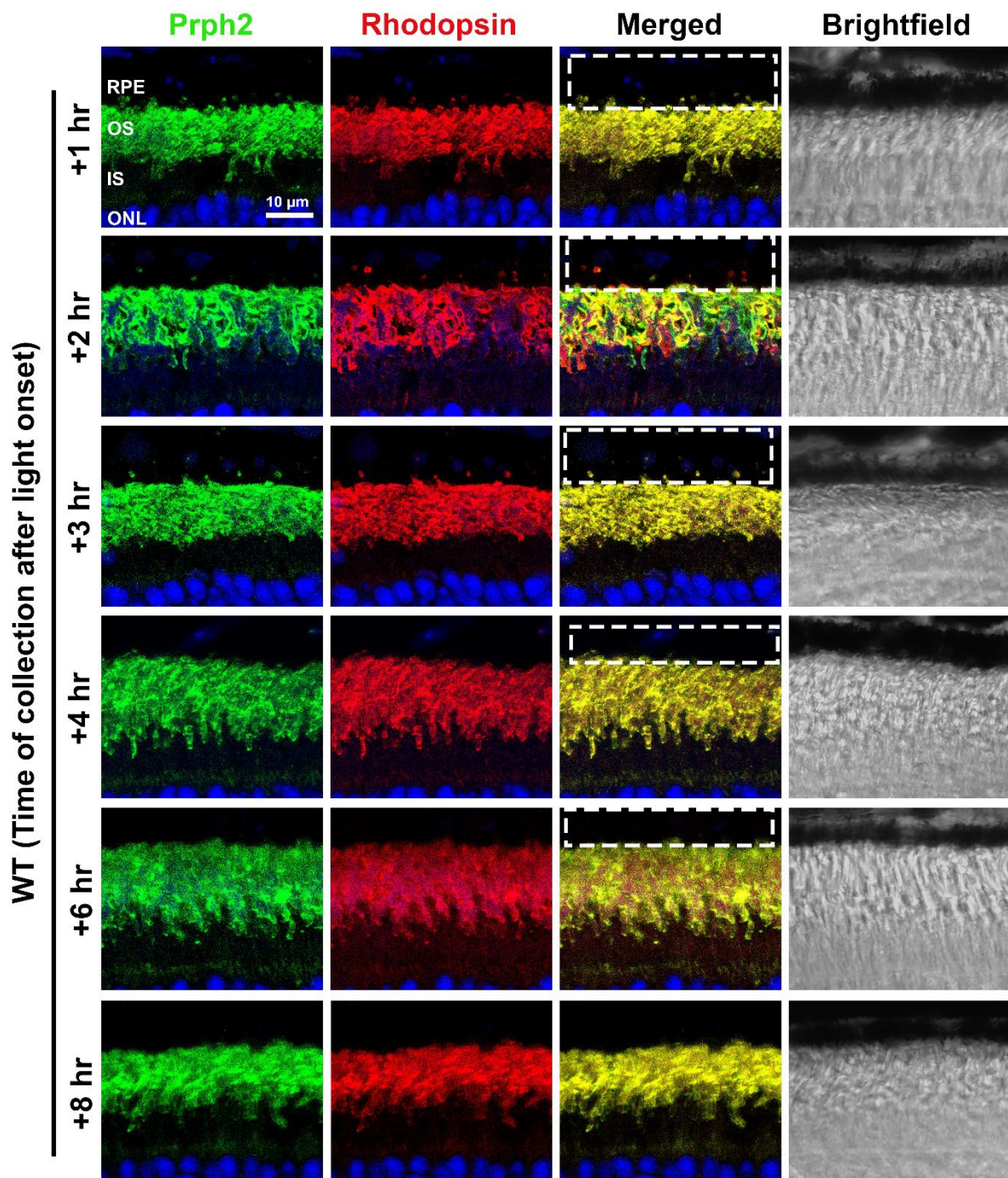


**Supplemental Figure 1. Structural and functional quantification of *Prph2* disease models.** A. Mitochondrial length was measured across multiple electron micrographs as shown in Figure 1. \*\*  $p < 0.01$  by one-way ANOVA with Tukey's post-hoc test. N B-C. ERG C-waves (from Figure 3) were normalized to their respective ERG A-wave values and are plotted. \*  $p < 0.05$  by one-way ANOVA with Tukey's post-hot test.



**Supplemental Figure 2. *Prph2* disease models decreased rod and cone function. A-B.** Full field ERGs were performed under scotopic and photopic conditions. **A.** Representative ERG waveforms from each genotype at P30 are shown. **B.** Plotted is maximum a- or b-wave amplitude measured from disease models at P30 (top) and P180 (bottom). (**Scotopic a-wave P30:** WT N=16;

*Prph2*<sup>+/-</sup> N=19; *Prph2*<sup>Y141C/+</sup> N=10; *Prph2*<sup>C213Y/+</sup> N=15; *Prph2*<sup>k153del/+</sup> N=15; *Prph2* R172W N=13.  
**Scotopic a-wave P180:** WT N=18; *Prph2*<sup>+/-</sup> N=16; *Prph2*<sup>Y141C/+</sup> N=21; *Prph2*<sup>C213Y/+</sup> N=28;  
*Prph2*<sup>k153del/+</sup> N=26; *Prph2* R172W N=5. **Scotopic b-wave P30:** WT N=16; *Prph2*<sup>+/-</sup> N=19;  
*Prph2*<sup>Y141C/+</sup> N=10; *Prph2*<sup>C213Y/+</sup> N=15; *Prph2*<sup>k153del/+</sup> N=15; *Prph2* R172W N=12. **Scotopic b-**  
**wave P180:** WT N=20; *Prph2*<sup>+/-</sup> N=21; *Prph2*<sup>Y141C/+</sup> N=21; *Prph2*<sup>C213Y/+</sup> N=28; *Prph2*<sup>k153del/+</sup>  
N=28; *Prph2* R172W N=5. **Photopic b-wave P30:** WT N=16; *Prph2*<sup>+/-</sup> N=19; *Prph2*<sup>Y141C/+</sup> N=10;  
*Prph2*<sup>C213Y/+</sup> N=13; *Prph2*<sup>k153del/+</sup> N=15; *Prph2* R172W N=13. **Photopic a-wave P180:** WT N=16;  
*Prph2*<sup>+/-</sup> N=17; *Prph2*<sup>Y141C/+</sup> N=21; *Prph2*<sup>C213Y/+</sup> N=29; *Prph2*<sup>k153del/+</sup> N=28; *Prph2* R172W N=4).  
Shown is mean ± SD, \*\* p<0.01, \*\*\* p<0.001, and \*\*\*\* p<0.0001 determined by two-way  
ANOVA, with Sidak's post-hoc test.



**Supplemental Figure 3. Time course of disc shedding, phagocytosis and degradation of the shed discs in WT mice.** P30 WT eyes were collected at 1-8 hours after light onset as indicated. Retinal cross sections were co-stained with Prph2 (RDS-CT, green) and rhodopsin (red) and counterstained with DAPI. Dashed boxes highlight the RPE layer. Magnification 63x. Scale

bars: 10  $\mu\text{m}$ . RPE: retinal pigment epithelium, OS: outer segments, IS: inner segments, ONL: outer nuclear layer.