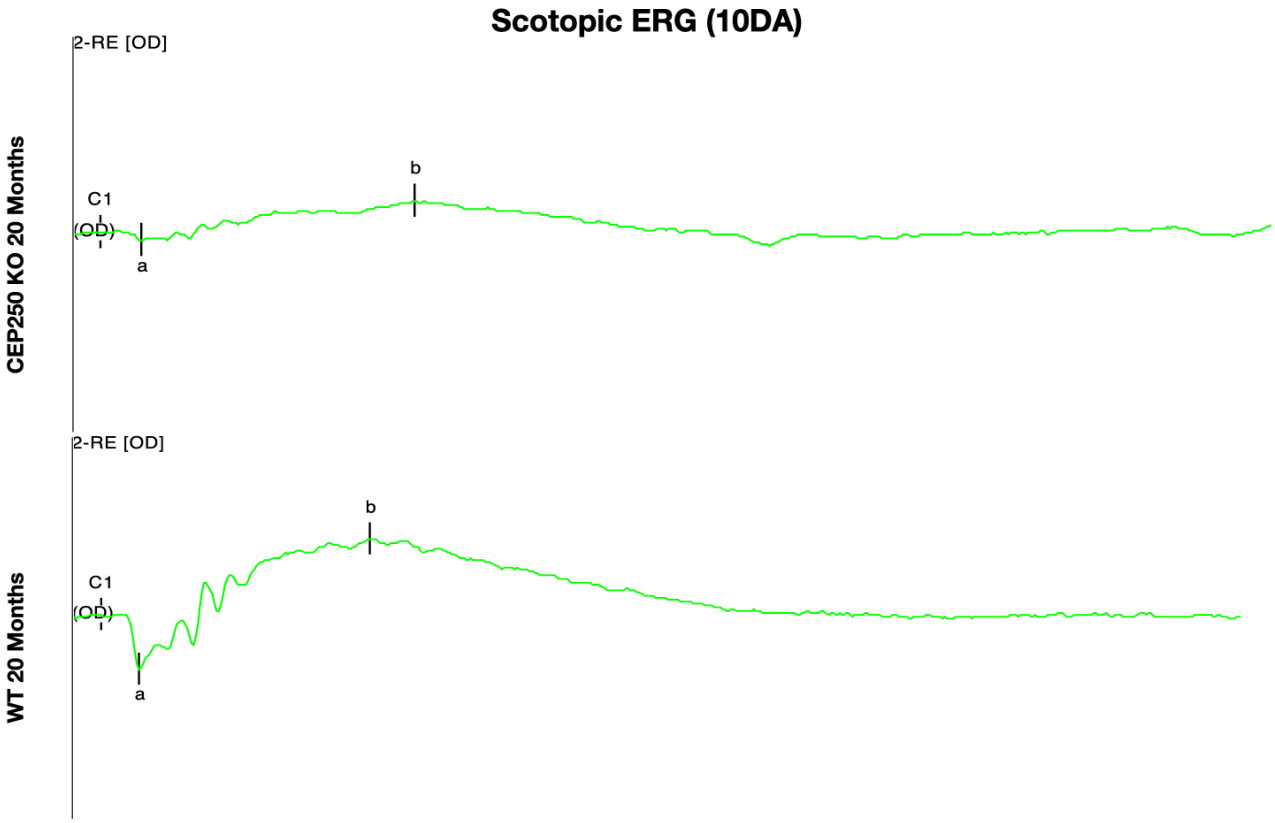
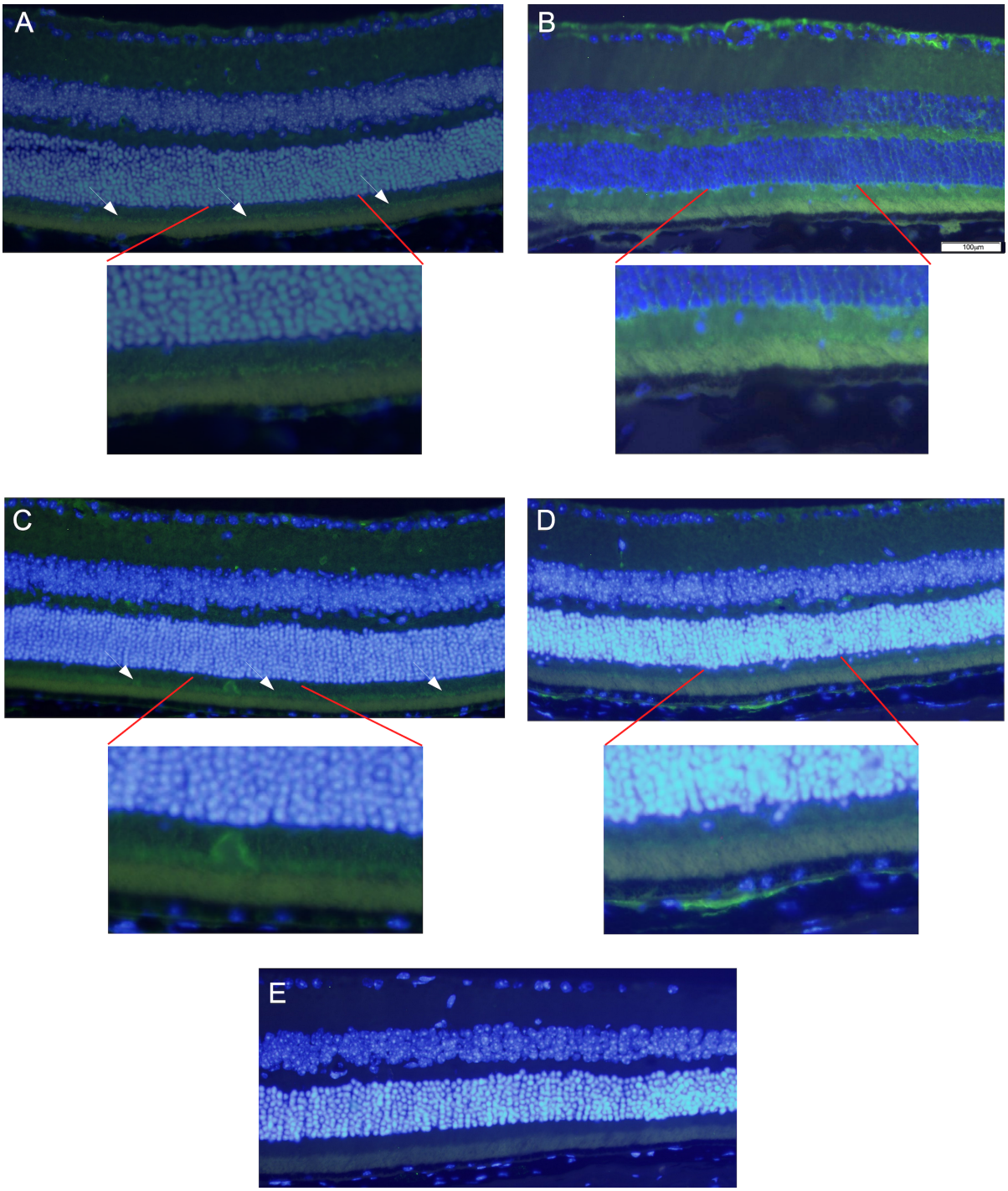
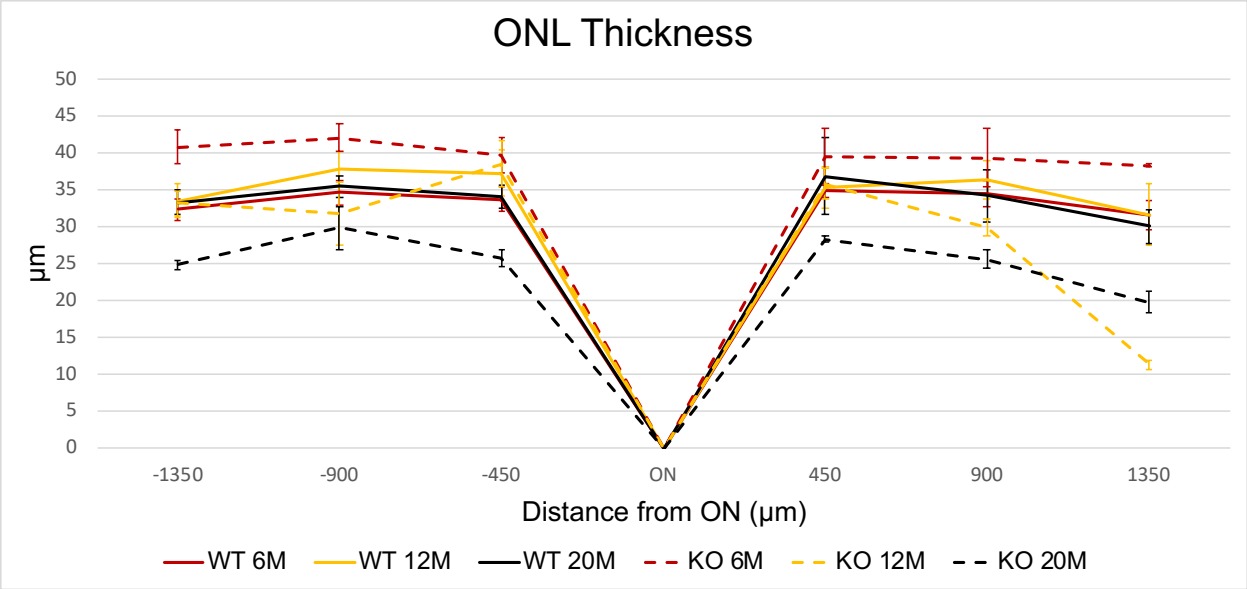


Supplementary Figure 1: Representative scotopic full field electroretinography (FFERG) of a CEP250 knock-out (KO) mouse at the age of 20 months (upper panel), compared to a wild type (WT) mouse at the age of 20 months (bottom panel) from the same background. The a-wave and b-wave amplitudes are lower in the KO mouse compared to WT indicating deterioration in retinal function due to the photoreceptor degeneration process.



Supplementary Figure 2: Immunohistochemical analysis of Cep78 (panels A and B) and Cep250 (panels C and D) in six months old WT (panels A and C) and Cep250-/- (panels B and D) retina. Both Cep78 and Cep250 are ciliary proteins and show expression in the junction between photoreceptor inner and outer segments (white arrows in panels A and C) in the WT retina. On the other hand, no Cep250 expression is evident in the Cep250 -/- retina (panel D) and diffused Cep78 staining is evident in the Cep250-/-. Panel E represents a negative control experiment with no primary antibody.



Supplementary Figure 3: Spider analysis of the ONL of WT and Cep250 KO retina at ages 6M, 12M, and 20M. Retinal sections were measures at six different locations along the retina in fixed distances from the optic nerve. The number of measurements varies depending on the section quality: WT samples: 6M n=6, 12M n=8, 20< n=4; KO samples: 6M n=4, 12M n=2, 20M n=2 and the presented data include average and standard deviation. The distance between the optic nerve and the far peripheral retina was measured from each side and divided into three fixed locations