Supplementary Information

Phagosomal and mitochondrial alterations in RPE may contribute to *KCNJ13* retinopathy

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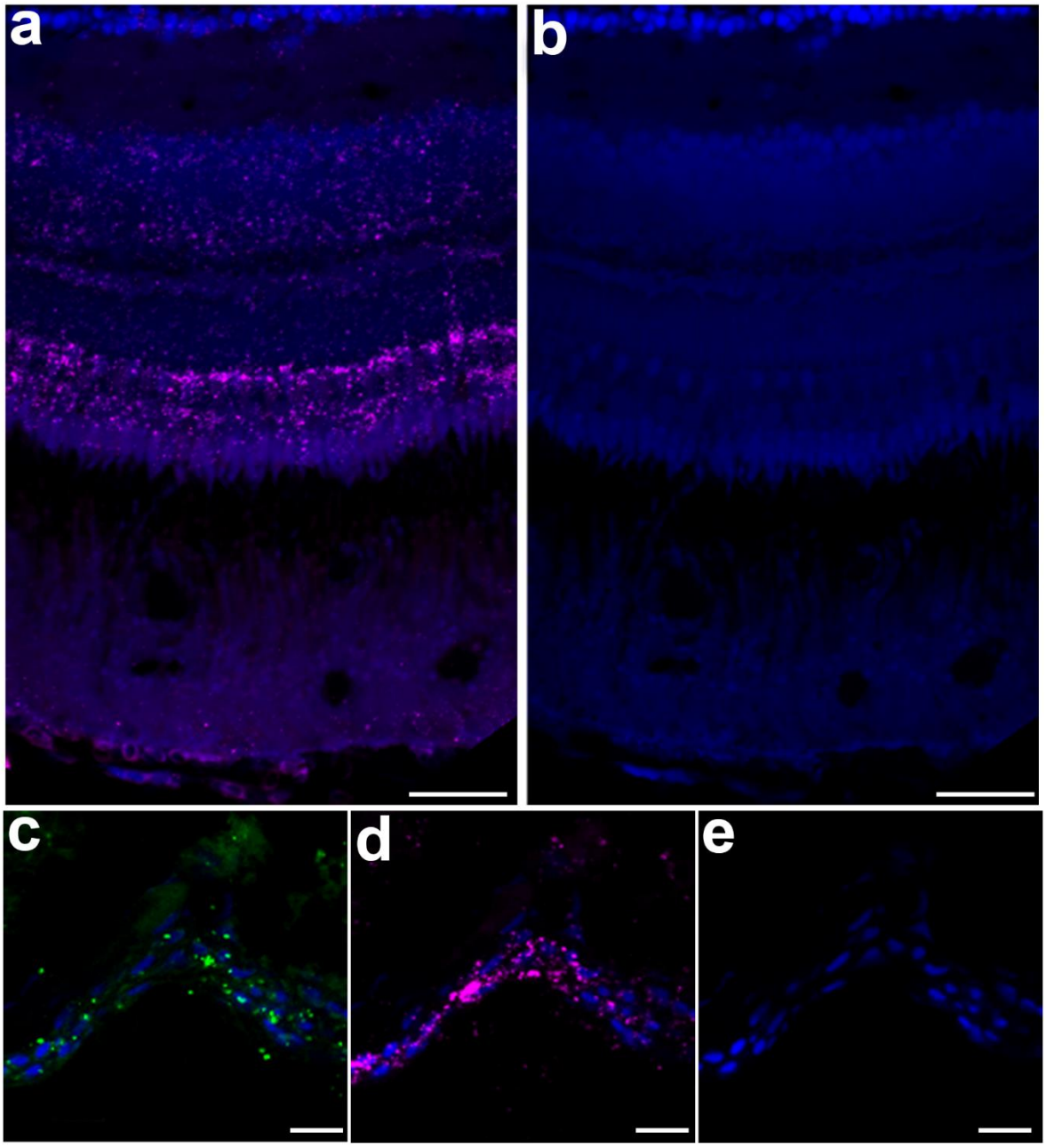
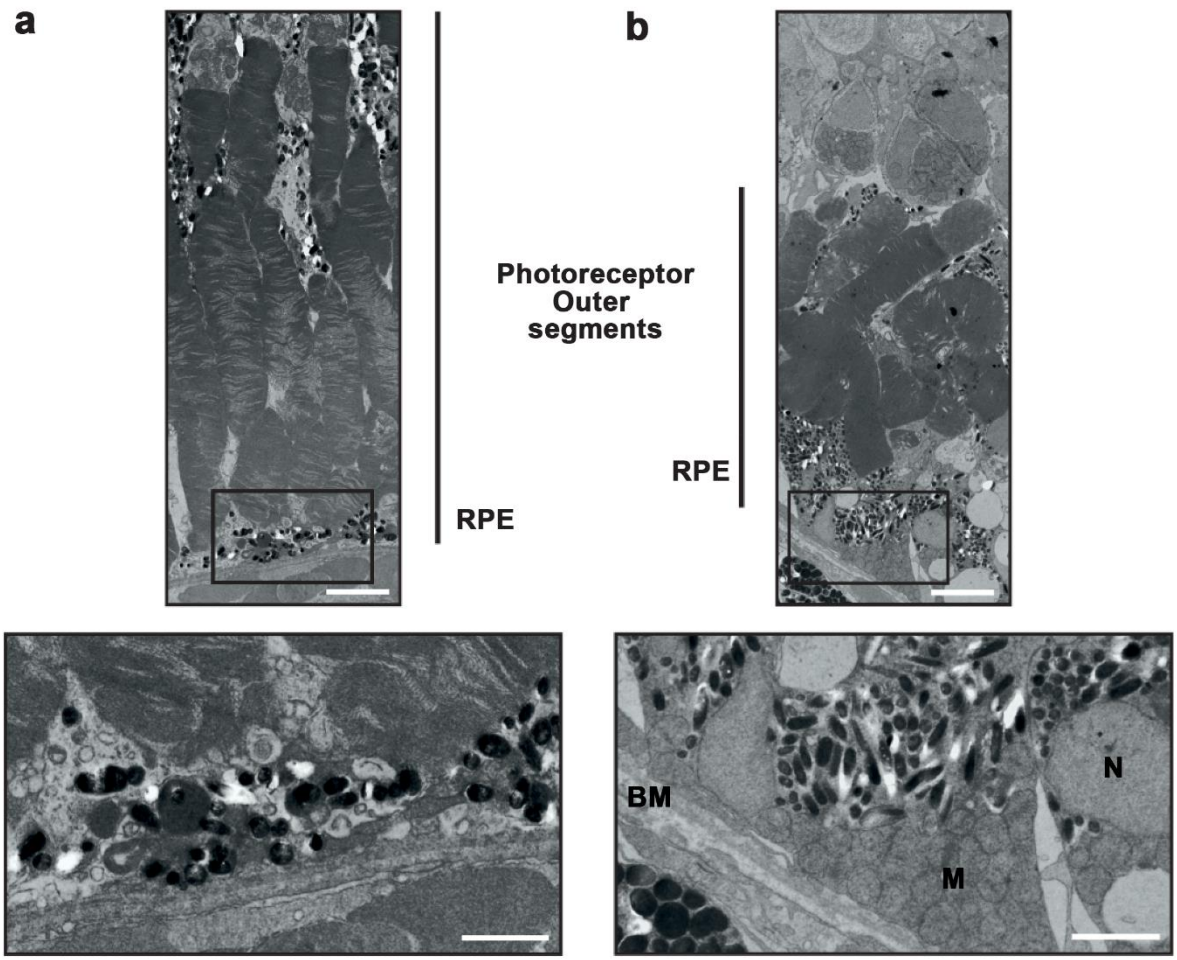


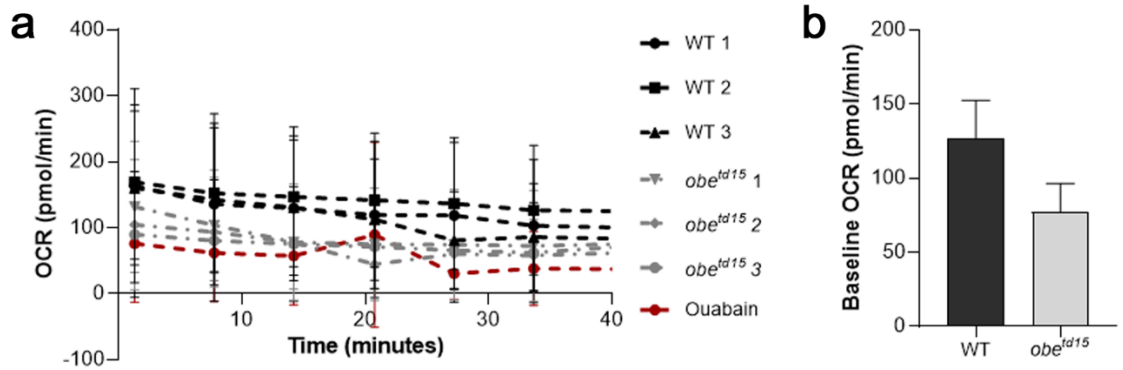
Supplementary Figure S1. RNAscope *in situ* hybridization positive and negative probes.

Wild-type adult retinal cryosections with *odc1* positive control probe (magenta) (a) and *dapB* (bacterial gene) negative control probe (b). Wild-type adult tail fin cryosections with *kcnj13* (green) (c), *odc1* (d) and *dapB* (e) probes. Sections are counterstained with DAPI (blue). Scale bars = 25 µm (a-b), 10 µm (c-e).



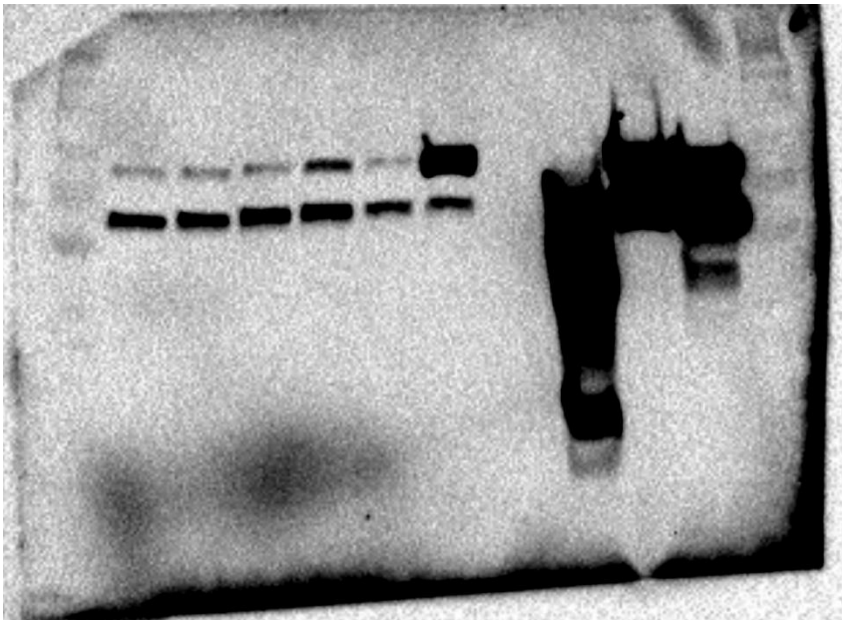
Supplementary Figure S2. *obe*^{td15} zebrafish eyes exhibit areas of photoreceptor loss with islands of preserved retina.

TEM images (a-b) of the photoreceptor outer segment and retinal pigment epithelium (RPE) from a 12 months post-fertilization *obe*^{td15} zebrafish eyes. A region displaying preserved retinal tissue (a) and an affected region (b) with disordered photoreceptor outer segments and numerous, large mitochondria in the RPE. BM, basement membrane; M, mitochondria; N, nucleus. Scale bars = 5 μm and 2 μm.



Supplementary Figure S3. Measurement of mitochondrial function using Seahorse XF analysis.

(a) Oxygen consumption rate (OCR) of 6 months post fertilization wild-type, *obe*^{td15} and ouabain-injected (4 days post injection) retinal samples (0.75 mm punch biopsies). n=6 per fish (3 punches taken per retina), mean ±SD. Baseline OCR is plotted on bar chart (b), p=0.0558. n=3, mean ±SD.



Supplementary Figure S4. Uncropped Western blot for hsp60