**Figure S1. Spider graphs showing the number of ONL nuclei and IS area at each part of the retina**
The number of ONL nuclei (A) and IS area (B) at each part of the retina were shown. Protective effects of PBA were observed in the superior retina as shown in the right side to the ON in the graphs (A, B). n for WT treated with vehicle, 3; P23H RP models treated with vehicle, 8–9; P23H treated with PBA, 8. ON, optic nerve. * p < 0.05, ** p < 0.01 for comparing P23H knock-in heter-ozygotes treated with vehicle and PBA, and ## p <0.01 for comparing WT and P23H knock-in heterozygotes treated with vehicle.
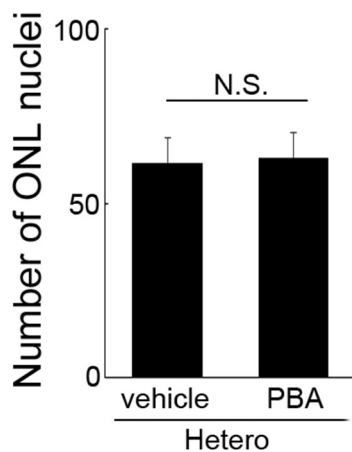
Figure S2. Number of photoreceptors in the P23H knock-in heterozygotes treated with PBA five times a week from 2 weeks until 4 weeks of age was comparable to those treated with vehicle
Data are shown as mean ± standard deviation. n = 6–8. p = 0.71. N.S., not significant.

Table S1. Primers for RT-PCR

gene (mouse)	Forward primer (5′–3′)	Reverse primer (3′–5′)
Gapdh	AGGAGCGAGACCCCACTAAC	GATGACCCTTTTGGCTCCAC
Rhodopsin	AACTTCGGCCCCATCTTCA	CAGTGGATTCTTGCCGCAG
Crx	TGGAGGAGCTGGAGGCCCTGTTTGCCAAGAC	CCAAAGGATCTGTACAAACATCTGTAGAG
xbp1s	CTGAGTCCGCAGCAGGTG	TGCCCAAAAGGATATCAGACT
Vcp	AAGTCCCCAGTTGCCAAGGATG	AGCCGATGGATTTGTCTGCCTC
Derl1	CGCGATTTAAGGCCTGTTAC	GGTAGCCAGCGGTACAAAAA
Fis1	ATATGCCTGGTGCCTGGTTC	AGTCCCGCTGTTCCTCTTTG
Mfn1	GATGTCACCACAGAGCTGGA	AGAGCCGCTCATTCACCTTA
Mfn2	CCCCTCTCAAGCACTTTGTC	ACCTGCTCTTCCGTGGTAAC
Pgc1a	GATGAATACCGCAAAGAGCA	AGATTTACGGTGCATTCCT
Tfam	AGTCAGCTGATGGGTATGGAGAA	TGCTGAACGAGGTCTTTTGG
Lc3b	Taqman probe; Mm00782868	

gene (human)	Forward primer (5′–3′)	Forward primer (5′–3′)
Gapdh	CACCCACTCCTCCACCTTT	TCCACCACCCTGTTGCTGTAG
Pgc1a	GCACCGAAATTCTCCCTTG	GCCTCTCGTGCTGATATTCC
Tfam	AAGATTCCAAGAAGCTAAGGGTGA	CAGAGTCAGACAGATTTTTCCAGTTT

Table S2. Primers for ChIP-qPCR

	Forward primer (5′–3′)	Reverse primer (3′–5′)
promoter gapdh	ACTAGGCGCTCACTGTTCTC	GTTGACTCCGACCTTCACCT
promoter pgc-1α		
probe 1	TAATAGCATCTGAGGGAAGCGTC	GTGCAACCAGGACTCTGAGT
probe 2	ACTCCAATCCACAGTGACACAG	GACAGGTGCCTTCAGTTCA
probe 3	AGCTCCCGAAGAGTTGCTG	GAAGGAAGCTGAAAGGATGGG
probe 4	AATGAGGGCTAATGCAGGTAGG	CCACTGTGTCCAGTACCTTGA
probe 5	AATCCAGCCTCATACCAGTCC	CTTCACTTGCTGCTTCTGGTC
probe 6	TGGGCAACAGAGTAAGACTCC	CCTCTCATTGTCCCAAGCTCAT
probe 7	CCCAGCAATGCACAACTCC	GGAGAATGATTGCACAGCAT