

SUPPLEMENTARY MATERIAL

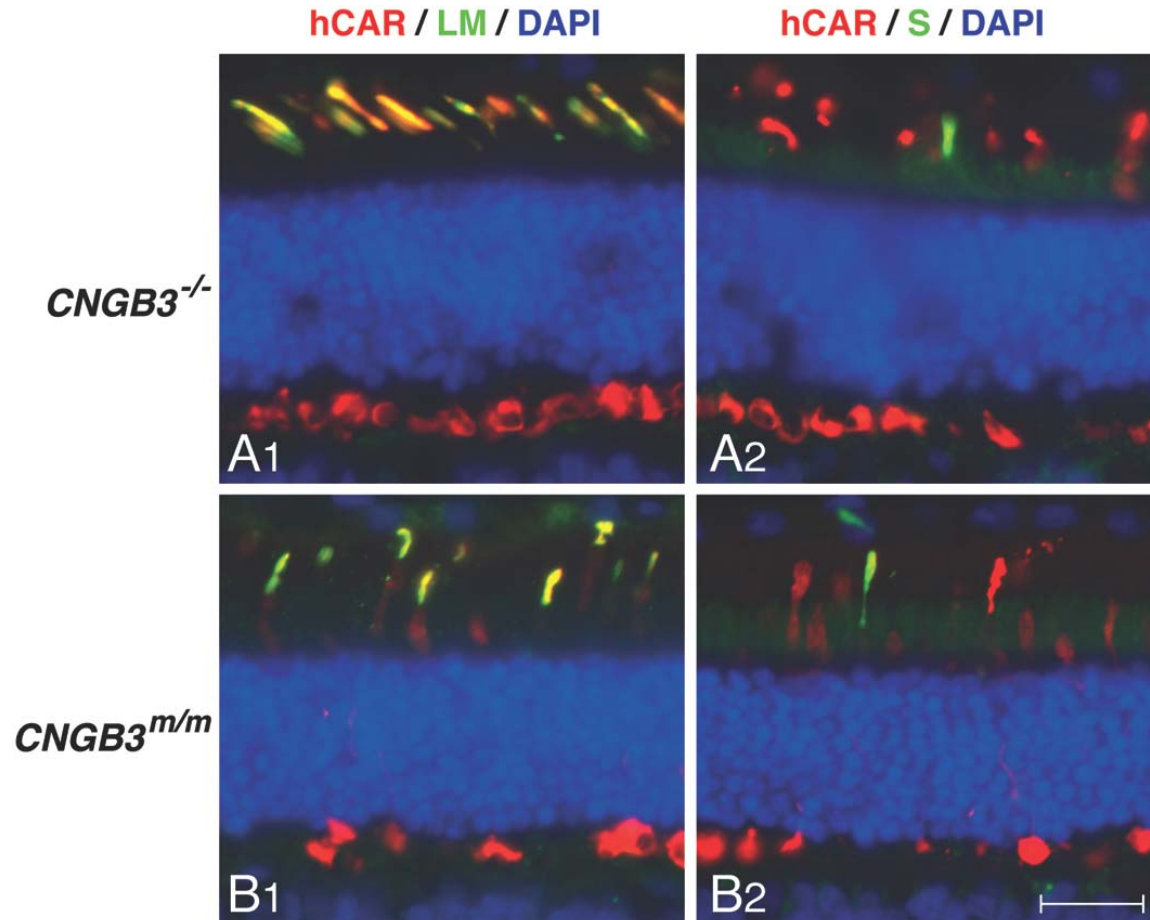


Figure S1. Cones are still present in older *CNGB3*-mutant canine retinas at several years of age. Shown are cones in a 1.1-year old *CNGB3*^{-/-} (A1, A2) and a 1.6-year old *CNGB3*^{m/m} dog (B1, B2). The cone outer segments are not as numerous and their morphology appears more irregular than in the younger *CNGB3*-mutant retinas shown in Figure 2. Calibration bar = 20 μ m.

Table S1. Primers and probes for qRT-PCR and exon junction crossed by PCR product.

| Gene | Forward Primer | MGB Probe | Reverse Primer | Exon Junction |
|------------------|------------------------------|----------------------|--------------------------|----------------------|
| <i>hCNGB3</i> | AAGAAGGCTCTCACCCAAGTAATC | TCAGCAAACCACAGCAC | GTGTGGCTCTTCAGACGTGACT | 1-2 |
| <i>cCNGA3</i> | GCCCTGCCTGTCTTCTATAACTG | TGTGCAGGGCCTGTT | CACAGCATCACGTGCTCAGA | 5-6 |
| <i>cCNGB3</i> | AAGATCCTGATCCAAGCAATCAG | CTCAGCAATCTACAAGACA | CTTCAAACGTGACTGGAGTCATCT | 1-2 |
| <i>L/M opsin</i> | CAGCGTCATCATACTGTGCTACCT | ATCCGAGCGGTGGC | GGACTCAGATTCTTTCTGCTGCTT | 4-5 |
| <i>S opsin</i> | CGCCATGTTTGTGCTTTGG | CTCTACAGCAGGTCTGGTGA | GATGACAATGTAGCGCTCAAAGG | 1-2 |
| <i>Rod opsin</i> | ATGATTGT CATATTCTTCTGCTATGGA | CAGTCAAGGAGGCAGCT | TGGGTGGTGGCCGATTC | 3-4 |

Movie S1. Behavioral visual performance of an untreated and treated achromatopsia affected dog under photopic conditions (sunny day).

- Part 1: Achromatopsia affected dog (*CNGB3^{m/m}*) – untreated.
- Part 2: Achromatopsia affected dog (*CNGB3^{m/m}*) – treated (GS80; see Table 1).

Two *CNGB3^{m/m}* mutant littermates are shown outside during a sunny summer day. The brown dog is untreated and shows the essential features of achromatopsia under photopic conditions, including disorientation, hypermetric gait, and collision with obstacles. The brown-grey, treated dog (grey-brown) acts visual with a normal gait; he is avoiding obstacles and finds toys based on visual cues.

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Movie S2. Behavioral visual performance of a normal control dog, as well as an untreated and treated achromatopsia affected dog in the obstacle avoidance course.

- Part 1: Normal control dog.
- Part 2: Achromatopsia affected dog (*CNGB3^{-/-}*) – untreated.
- Part 3: Achromatopsia affected dog (*CNGB3^{-/-}*) – treated (M626; see Table 1).

The movies show a normal control dog as well as an untreated and a treated *CNGB3*-mutant dog