SUPPLEMENTARY MATERIAL

Figure S1 Electron micrographs of normal canine retinas 1 week following intravitreal injection of PBS or CNTF. Compared to the PBS control retina (a), the CNTF treated retina shows severe shortening and disorganization of the rod and cone photoreceptor outer segments (b1, b2). Cone inner segments are labeled (**C**). Calibration bars = 2.5 µm.

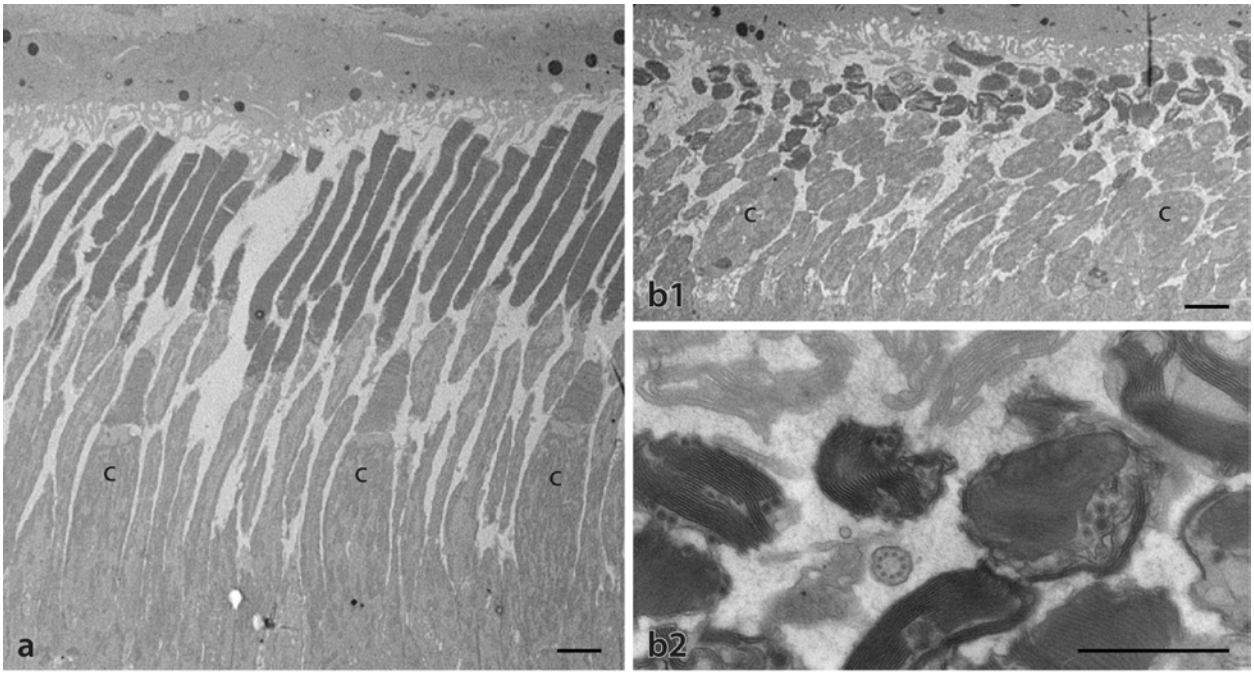

Figure S1

Figure S2 Partial restoration of CNGA3 and GNAT2 protein localization in the *CNGB3*-mutant retina 1 week following intravitreal CNTF injection. Normal (wt) cone photoreceptors show both CNGA3 (A2) and GNAT2 (A5) present in the outer segments and co-localize with L/M-opsin (A1, A3 and A4, A6). The cone outer segments of untreated, adult mutants [ACHM (untreated)] contain no detectable CNGA3 (B2) and GNAT2 (B5). L/M-opsin localization remains unaffected by the mutation (B1, B3, and B4, B6). One week following intravitreal CNTF in the mutant eye [ACHM (1 week post CNTF)] cone outer segments are fewer in numbers and severely shortened (C1, C4), but some of them weakly express CNGA3 (arrows in C2 and C3) and GNAT2 (arrows in C5 and C6). Cell nuclei are shown in blue with DAPI. ACHM, achromatopsia-affected; wt, wildtype. Calibration bar = 10 µm.

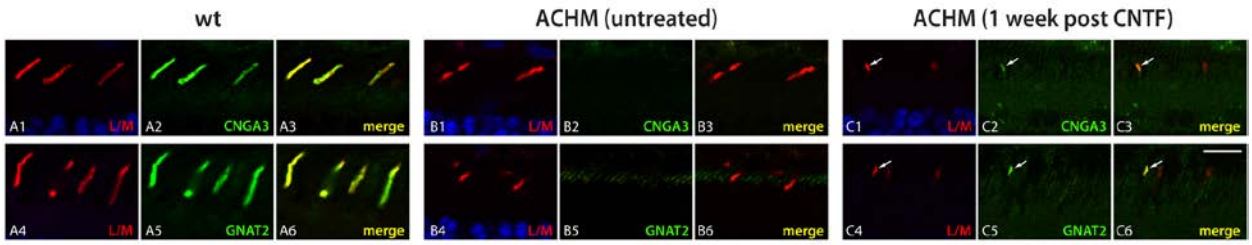

Figure S2

Table S1. Intravitreal CNTF injections in normal dogs.

Dog ID / gender	Age (months)	Eye	Treatment	Studies (weeks post treatment)				
				ERG	IHC	EM	qRT-PCR	Western
M636 / m	7.3	R	CNTF	0, 1	1			
		L	PBS	0, 1	1			
M637 / m	7.3	R	CNTF	0, 1				
		L	PBS	0, 1				
M638 / m	7.3	R	CNTF	0, 1			1	
		L	PBS	0, 1			1	
M670 / m	4.5	R	CNTF	0, 0.5, 1				1
		L	PBS	0, 0.5, 1				1
M667 / m	5.8	R	CNTF	0, 1				
		L	PBS	0, 1				
M645 / m	19.8	R	CNTF	0, 1		1		
		L	PBS	0, 1		1		
GS122 / m	6.3	R	CNTF	0, 2	2			
		L	PBS	0, 2	2			
GS125 / f	6.3	R	CNTF	0, 2			2	
		L	PBS	0, 2			2	
M672 / f	4.5	R	CNTF	0, 0.5, 2				2
		L	PBS	0, 0.5, 2				2
M640 / f	7.3	R	CNTF	0, 5			5	
		L	PBS	0, 5			5	
M641 / f	7.3	R	CNTF	0, 5	5			
		L	PBS	0, 5	5			
GS121 / m	6.3	R	CNTF	0, 5				5
		L	PBS	0, 5				5

Gender: m, male; f, female
Eye: R, right; L, left
Treatments: Intravitreal injection of either CNTF (12 µg in 30 µL PBS) or PBS (30 µL).
Studies: ERG, electroretinogram; IHC, immunohistochemistry; EM, electron microscopy; qRT-PCR, quantitative real-time PCR; Western, western blot

Table S2. Antibodies and reagents used for immunohistochemistry (IHC) and western blotting (WB).

Antigen	Expected retinal localization or target protein	Host	Working dilution	Source, Catalog Number or Name
Human cone arrestin	Cone photoreceptors	Rabbit polyclonal	1 : 10,000	Cheryl Craft (University of Southern California)
L/M-cone opsin	Outer segments of L/M-cones	Rabbit polyclonal	1 : 100	Chemicon, AB5405
		Goat polyclonal	1 : 100	Santa Cruz, sc-22117
S-cone opsin	Outer segments of S-cones	Rabbit polyclonal	1 : 5,000	Chemicon, AB5407
		Goat polyclonal	1 : 50	Santa Cruz, sc-14363
Cone *alpha* transducin (GNAT2)	Cone outer segments	Rabbit polyclonal	1 : 5,000	Santa Cruz, sc-390
Alpha subunit of cone cyclic nucleotide-gated channel (CNGA3)	Cone outer segments	Rabbit polyclonal	1 : 5,000	Komáromy and colleagues[10]
Rod opsin	Rod outer segments	Mouse monoclonal	1 : 100	Chemicon, MAB5316
pSTAT1 (Tyr701)	Activation of Jak/Stat signaling pathway	Rabbit monoclonal	1 : 1,000 (IHC) 1 : 2,000 (WB)	Cell Signaling Technology, #9167
pSTAT3 (Tyr705)	Activation of Jak/Stat signaling pathway	Rabbit polyclonal	1 : 1,000 (IHC) 1 : 2,000 (WB)	Cell Signaling Technology, #9131
Phospho Histone H3 (Ser 10) (PHH3)	Cell mitosis	Rabbit polyclonal	1 : 50	Millipore, 06-570
TUNEL (In Situ Cell Death Detection Kit, Fluorescein)	Apoptotic cell nuclei		kit instructions	Roche, 11684 795 910
Actin	Loading control (WB)	Mouse Monoclonal	1 : 20,000	Chemicon, MAB1501

Chemicon, Millipore Corporation, Temecula, CA; Santa Cruz Biotechnology Inc., Santa Cruz, CA; Cell Signaling Technology Inc., Danvers, MA; Roche Applied Science, Indianapolis, IN.

Table S3: Designed primers and probes for qRT-PCR and exon junction crossed by PCR product.[10]

Gene	Forward Primer	MGB Probe	Reverse Primer	Exon Junction
hCNGB3	AAGAAGGCTCTCACCCAAGTAATC	TCAGCAAACCACAGCAC	GTGTGGCTCTTCAGACGTGACT	1-2
cCNGA3	GCCCTGCCTGTCTTCTATAACTG	TGTGCAGGGCCTGTT	CACAGCATCACGTGCTCAGA	5-6
cCNGB3	AAGATCCTGATCCAAGCAATCAG	CTCAGCAATCTACAAGACA	CTTCAAACGTGACTGGAGTCATCT	1-2
cL/M opsin	CAGCGTCATCATACTGTGCTACCT	ATCCGAGCGGTGGC	GGACTCAGATTCTTTCTGCTGCTT	4-5
cS opsin	CGCCATGTTTGTGCTTTGG	CTCTACAGCAGGTCTGGTGA	GATGACAATGTAGCGCTCAAAGG	1-2
cCNGA1	ATGAAACCCCCCATGCAA	ATCCATCACAGAGGGA	TGGCACCAGGCAGGTACTG	2-3
cCNGB1	TGCATTATCCTGAGTCTCAGAAGTTAC	AGAAGGCCAGGCGCA	TCCTTGGGCTTGTTGTTATTTCTC	16-17
cRho	ATGATTGTCATATTCTTCTGCTATGGA	CAGTCAAGGAGGCAGCT	TGGGTGGTGGCCGATTC	3-4