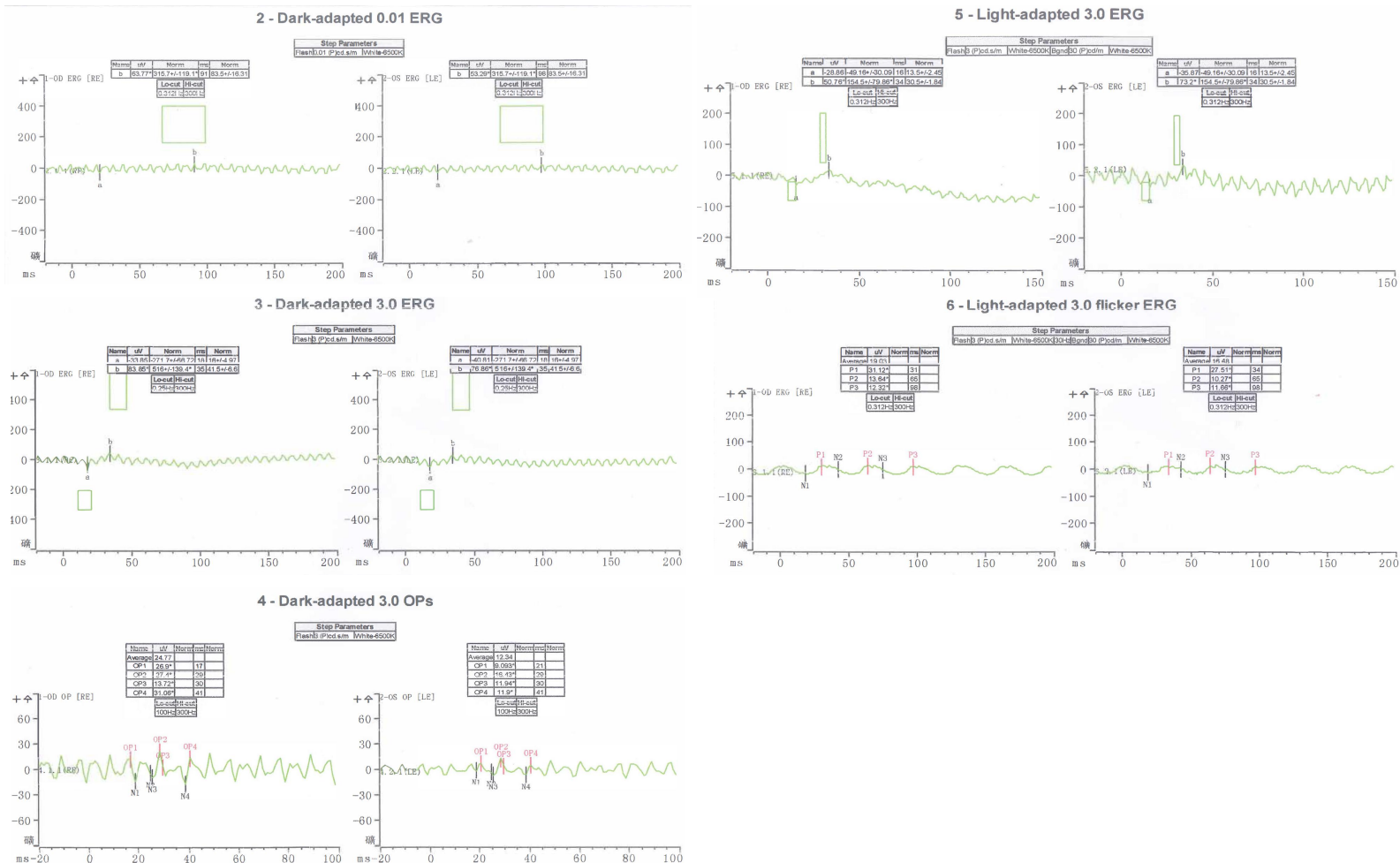




Supplementary Figure S1. Electrophysiological Assessment of Retinal Function in Proband 1.

Electroretinogram (ERG) recordings from Proband 1 at Changsha Aier Eye Hospital on 05-16-2021, demonstrating severely impaired retinal function consistent with Leber congenital amaurosis (LCA). The test protocol followed the ISCEV Standard (2008).

Panels 2 & 3 (Dark-adapted ERG): Show markedly reduced or absent responses to both low-intensity (0.01 cd.s/m²) and high-intensity (3.0 cd.s/m²) flashes under dark-adapted conditions, indicating profound rod system dysfunction.

Panel 4 (Dark-adapted OPs): Reveals a near-complete absence of oscillatory potentials (OPs), which are generated by inner retinal neurons (primarily amacrine cells), suggesting secondary involvement of the inner retina.

Panels 5 & 6 (Light-adapted ERG): Display severely attenuated responses to single flash (3.0 cd.s/m²) and flicker (30 Hz) stimuli under light-adapted conditions, reflecting significant cone system impairment.

These findings corroborate the clinical diagnosis of LCA and provide objective evidence of widespread photoreceptor and inner retinal dysfunction in the patient.