

Supplementary Figure S7. Electroretinography from the right eye of patients affected by homozygous c.498_499 mutations and a representative normal subject. Patients 1 and 2 demonstrate a complete loss of electrophysiological responses. The ffERG from patient 3 demonstrates a loss of response under scotopic conditions. The cone response is delayed and is approximately 85% of control response. The combined and cone flicker in patient 3 show moderately attenuated responses, which are approximately 40% of normal with delayed responses. The scotopic responses from patient 4 are completely attenuated. The combined and cone flicker responses are severely reduced to 10% of normal.